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CONGENITAL DEFECTS IN THE PELVIC PARASYMPATHETIC SYSTEM*

BY

ORVAR SWENSON

From the Department of Surgery of the Boston Floating Hospital and Tufts College Medical School

Nine years ago, while observing a group of patients with congenital megacolon, three children came under my observation who had transverse colostomy. This vent in the colon had relieved the patients' symptoms completely. The colostomy was their only remaining problem. After discussing the patients with my colleagues, it was decided that the colostomies should be closed and medical therapy instituted. This programme was followed, and within seven months all of the patients' symptoms recurred despite careful medical management. Re-opening the colostomies again relieved all of their symptoms.

In reviewing this experience, it seemed obvious that such patients must have some form of functional obstruction in the colon beyond the colostomy. Careful barium enemas were performed on other patients with congenital megacolon, and in the distal portion of the large intestine there was a persistent narrowing of the lumen beyond the dilated colon which varied in length from patient to patient, but otherwise was quite similar. Usually the rectum and rectosigmoid and varying parts of the sigmoid colon were irregular and narrow in calibre, and the colon above this narrow portion was considerably dilated (Fig. 1).

In the course of performing transverse colostomies on patients with Hirschsprung's disease, explorations were performed and the lesion in the distal colon was examined. The dilated left colon was markedly hypertrophied and within a distance of 5 to 8 cm. the calibre of the bowel abruptly narrowed so that the lumen became smaller in diameter than normal. The narrow segment was little if at all hypertrophied.

A group of patients with transverse colostomies was accumulated, and these afforded an opportunity to study the motility of the left colon. A multiple balloon technique was used in which one balloon was located in the splenic flexure, one in the descending colon, and one in the narrow rectosigmoid. A number of normal individuals were studied, and it was found that in the normal intact left colon peristaltic waves progressed in an orderly fashion from the splenic flexure through the rectum. Patients with megacolon exhibited marked variations from the normal. The peristaltic waves progressed from the splenic flexure to the descending colon in an orderly manner. However, the balloon in the rectosigmoid failed to record any progression of the peristaltic waves into the narrow portion of the colon. There were frequent, feeble, purposeless contractions which were unrelated completely to the normal peristaltic waves in the dilated colon above (Swenson, Rheinlander and Diamond, 1949). These studies led to the conclusion that there was a segment

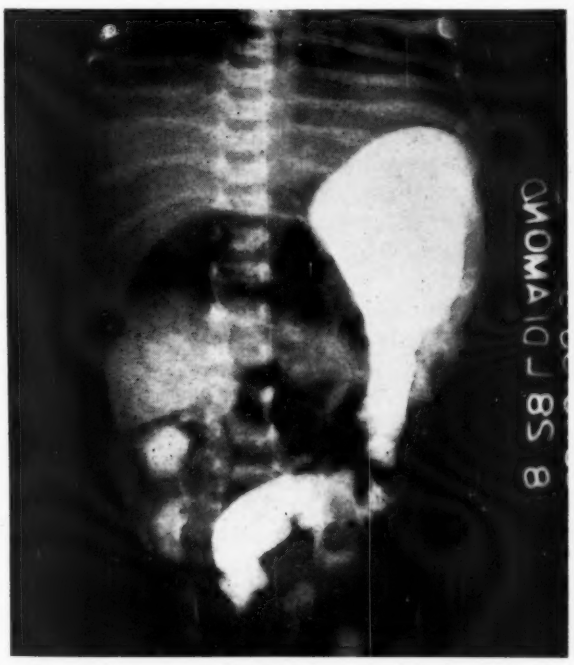

FIG. 1.—Appearance of a barium enema in a patient with Hirschsprung's disease. This lesion was higher than usual, being in the splenic flexure.

* The 1954 Alex Simpson-Smith Memorial Lecture delivered at the inaugural meeting of the British Association of Paediatric Surgeons at The Hospital for Sick Children, London.

of colon in these patients which was without peristaltic activity. It was tempting to postulate the presence of spasm in the colon beyond the dilatation, and there was some evidence that this was true. However, it was not possible to prove this point definitely by direct measurements.

It was concluded from these studies, as well as from our observation of the patients with colostomy, that if the inactive segment of bowel were resected and continuity of the intestinal tract were restored, these patients should be relieved of their functional obstruction. In the laboratory a technique was tested which consisted of resection of the recto-sigmoid and rectum with an anastomosis between the anal canal $1\frac{1}{2}$ cm. from the mucocutaneous margin and the colon above. Dogs were used in these studies, and it was found that the technical procedure was feasible and that the animals were continent post-operatively. This technique was utilized on patients and the results were most gratifying (Swenson and Bill, 1948).

Clinicians were slow to realize the significance of the absence of ganglion cells which had been described by a number of workers (Dalla Valle, 1920; Robertson and Kernohan, 1938; Tiffin, Chandler and Faber, 1940; Zuelzer and Wilson, 1948). In this series of 150 cases absence of ganglion cells has been a constant finding. The absence of ganglion cells extended to the internal sphincter in all except one of the specimens removed at operation in this series. This patient had a cuff of distal rectum 4-6 cm. long with normal ganglion cells and proximal to this a segment in which the ganglion cells were absent. Our experience has been that there is a total absence of ganglion cells except in the transition zone adjacent to the normal colon. Here, in a band 2-3 cm. in length, there is a decrease in concentration of ganglion cells.

In a review of this group of 150 patients it was found that the most important point in the history was the duration of the disease. Virtually all of the records revealed a history of symptoms dating from birth. This is in contrast to the children with habit constipation where the onset of symptoms is usually at 2 or 3 years of age. On examination of these patients, the finding of an impaction in the sigmoid and no impaction in the rectum is strong evidence of Hirschsprung's disease. Barium enema must be used to confirm the diagnosis of Hirschsprung's disease, and in the majority of patients the diagnosis can be made unequivocally.

In the past few years I have become increasingly aware of the difficulty in diagnosing Hirschsprung's disease accurately in newborn babies and infants under 6 to 8 months of age, for in this age group the barium enema often fails in making a definite diagnosis. Since the aganglionic segment in most patients extends down to the internal sphincter, we determined to biopsy the rectal wall through the anal canal in an attempt to develop a satisfactory biopsy technique for positive diagnosis of Hirschsprung's disease. Such a biopsy would also be useful in older patients who had equivocal signs of the disease on radiological examination. A third group of patients presented themselves who posed a diagnostic problem. These were the children who had had a transverse colostomy early in life. Whether or not these children had Hirschsprung's disease was important in their management. Placing barium in the deactivated colon distal to the colostomy was of no avail because this segment had become uniformly small in calibre. In order to test the validity of our biopsy technique, we performed the procedure on 10 cadavers. We learned that the internal sphincter itself was hardly a reliable source of biopsy material, but that immediately above this in the rectal wall a sufficient amount of rectal wall could be removed to make the diagnosis.

In this series of 10 cadavers we removed a triangular piece of rectal wall about 1 cm. in length and $\frac{1}{2}$ cm. in width. All of these proved to have ganglion cells. We have subsequently biopsied a group of 19 patients who at operation proved to have Hirschsprung's disease. In all of these no ganglion cells could be found in the biopsy material. This test has proved to be of considerable value in the three groups of patients which I have enumerated. An adequate sample of the rectal wall must be removed. We have now performed the biopsy on a total of 40 patients, and in none has there been any complication except bleeding in one patient post-operatively. Of the total number, 19 were aganglionic and proved to have Hirschsprung's disease; 21 had ganglion cells in the biopsy and these patients had histories and radiological findings consistent with chronic constipation. This seems a satisfactory and helpful technique in the various diagnostic problems with which one might be confronted in considering the diagnosis of congenital megacolon.

There are several points in the operative treatment of these patients that should be stressed. In the first place we are now resecting 75% of our patients without a preliminary colostomy. The multiple stage method of management is reserved for those patients who are extremely ill and emaciated. Colostomy is most frequently used in infants, for in our experience more severely ill patients are encountered in this age group than in any other. The colostomy should be performed in the left colon at the level of the junction of the aganglionic and

normal colon. This can be determined by performing a biopsy of the muscular coat at the proposed site of the colostomy. The bowel wall is incised down to the mucosa and the muscular coat freed for 4 to 5 mm. and a full thickness of muscular wall excised. This is immediately studied by frozen section and if the ganglion cells are found the colostomy is performed at this site. If none are found, the colostomy is placed at a higher level where the neuromuscular structures are intact. The advantage of a left-sided colostomy at the junction of the aganglionic and normal colon is that at the second procedure the colostomy can be resected along with the aganglionic colon and continuity of the intestinal tract established. Thus the programme is reduced to two operative procedures. Formerly, when the colostomy was performed in the transverse colon, we were committed to three operations: colostomy, resection of the aganglionic segment of colon with anastomosis, and then closure of the colostomy.

Preparation of these patients for operation is less elaborate than formerly. The parents are instructed to administer daily enemas to the child for three days preceding admission to hospital. No chemotherapy is used to prepare the intestinal tract. It is possible to be less vigorous in the preparation of these patients when a segment of the colon is resected during the abdominal phase of the operation. Large faecal impactions can be removed in this manner. If one utilizes the telescoping type of procedure in which there is no resection of bowel during the abdominal phase of the operation, a more elaborate preparation of the gastro-intestinal tract must be performed. I used the telescoping operation in my first few patients and found that it was so clumsy that it was discarded for the type of procedure now used. There are additional advantages in the resection type of procedure. A biopsy is secured from the proximal end of the resected bowel, and this ensures that the whole aganglionic segment has been removed. Another often unappreciated advantage is that one has a very small length of rectum and rectosigmoid to pull down through the anal canal. This is much neater and cleaner than everting the whole dilated intestine. The greatest advantage of all is that should one pull the short segment of bowel down and find that it has not been freed low enough, the small segment can be returned to the pelvis with little difficulty and the dissection continued. If the whole segment has been telescoped, it is an arduous task to reduce the intussusception to permit a completed dissection.

The second technical feature of the operative procedure that should be stressed is that in order to ensure a good blood supply at the proximal line of resection, it is advisable to free the splenic flexure and to divide the left colic artery. A more limited resection was performed early in my experience, and at times the blood supply was precarious in the proximal segment.

A few patients have been encountered with extensive lesions, some of which have extended to the transverse colon and even to the hepatic flexure. In these patients we had divided the middle colic artery and vein and rotated the ascending colon 180° down into the pelvis, and performed the reconstruction in this manner. It is surprising how well these patients have functioned post-operatively.

I have encountered two infants with the entire colon aganglionic, and the management of these babies was problematical. Initially both of these infants had ileostomies closed. An experience six years ago with a patient suggest a method of managing these two infants. This boy had a colectomy with an anastomosis between the ileum and rectosigmoid. With the short aganglionic segment in the pelvis, the proximal ileum dilated to enormous size, abdominal distension was massive, and the child was critically ill. It was not until ileostomy was performed that he began to gain and do well. Again in this boy the problem was what to do with the ileostomy. After considerable discussion with colleagues and with the parents, the rectum and rectosigmoid were resected and the terminal ileum anastomosed to the anal canal, utilizing the same technique of anastomosis as is used with the routine patients with Hirschsprung's disease. It took some time for the boy to adjust to the situation and he did have some rather hectic times. Gradually, over a period of months, he improved. He is now six years from operation and has grown and developed perfectly normally. He has no further troubles and only a moderate amount of dilatation of the terminal ileum. He has had two or three semi-formed evacuations daily; he is perfectly continent. On the basis of this experience it was decided that the same procedure should be performed on these two infants. Surprisingly their course was less complicated than the older child's had been. Immediately post-operatively they had as many as 15 loose stools a day, and the perineal care was a constant nursing problem for a matter of several weeks. Gradually the stools became firmer and movements were reduced to three or four a day. These two infants are now one and a half years and one year from operation, and their growth and development have been normal (Fig. 2). Barium enemas show some dilatation of the terminal ileum but far less than had been anticipated.

The next important technical detail of the operative procedure is the anastomosis. It must be made water-tight or leakage and pelvic infection may occur. First the muscular coats of the two segments are approximated with interrupted 00000 silk. This row must be placed completely around the bowel, and all the sutures tied before the proximal segment is opened. Thus, the first layer is made under aseptic conditions and the pelvis is protected from contamination by this suture line. When it has been completed, the proximal end is opened and the mucosa of the two segments are then approximated with interrupted chromic catgut sutures.

In our series of 150 patients, we have had three patients who have had leaks at the anastomosis. One of these was a patient who had such a short segment of bowel proximally that we could not complete the anastomosis, and we realized that some pelvic infection would be inevitable. The infection was surprisingly limited; however, this child developed a severe stricture which has been the source of chronic trouble. We have had five other minor strictures, all of which have yielded to digital dilatation. The second patient with infection had a perirectal abscess which was easily drained. Fortunately a fistula did not develop. The third patient with infection developed a rectovaginal fistula. This was probably due to a suture placed too deeply so that it perforated the vaginal wall. In this particular patient a small fistula developed, and a transverse colostomy was performed and she was left with a diverted faecal stream for six months. The colostomy was closed two years ago, and the fistula has not recurred. No operative procedure was performed in the region of the fistula.

FIG. 2.—Photograph of child whose entire colon was aganglionic. This was taken one and a half years after colectomy and anastomosis of the terminal ileum to the anal canal. This child is continent and growth and development have been normal.

Post-operatively our patients have done uniformly well and have been relieved of their symptoms. It has not been necessary to resort to laxatives or enemas in any of our patients post-operatively, except in three children who for a short time developed mild constipation at the age of 2 or 3 years. All occurred some months after operation and all are doing well now.

In this series, 20 patients were operated upon in the first six months of life, and in this group there were four post-operative deaths. Of the four, one had a multiple stage procedure and three had primary resections. In the remaining 130 patients, there was one post-operative death. This $2\frac{1}{2}$-year-old boy died with generalized convulsions, probably due to pulmonary infection and septicaemia. The abdomen and pelvis were free of infection. It is questionable whether the outcome would have been more favourable if the three with primary resections who died had been treated with preliminary colostomies. Of these three, the causes of death were intestinal obstruction, recurrence of the disease because of incomplete resection and bilateral adrenal haemorrhage. The patient with the multiple stage procedure had congenital heart disease.

There is one complication to which these patients seem to be prone which is worthy of careful consideration. For some obscure reason, some of these children with Hirschsprung's disease do not tolerate infection in a normal manner. This is particularly true of the so-called gastro-enteritis which we consider to be an intestinal infection of viral aetiology. A normal child with gastro-enteritis has a short illness with vomiting and diarrhoea. Some children with megacolon either pre- or post-operatively become desperately ill under similar conditions. Initially there may be vomiting and a few diarrhoea stools. Later progressive abdominal distension and obstipation develop. Profound dehydration occurs from loss of fluid into the gastro-intestinal tract as well as from the vomiting and diarrhoea. Part of the derangement of function is spasm of the sphincter mechanism so that nothing is released from the intestinal tract. This disease is most commonly seen in small infants. Treatment must be directed along two lines; first, dehydration must be combated, and as this is often greater

than anticipated, generous parenteral replacement must be provided. Second, a large soft rectal tube must be inserted through the anal canal, and the colon drained and gently irrigated with warm normal saline. Irrigation must be repeated three times a day. Usually in three or four days the child is perfectly well, and the stools become normal in consistency, and the children return to their usual habit of one or two evacuations daily. Most children with Hirschsprung's disease never have these attacks. Some seem to be more prone to them and to have them repeatedly. However, the tendency is that the attacks are less severe and less frequent as the child grows older.

Faecal incontinence has not occurred in our patients except for some soiling immediately post-operatively. All have been continent at the time of discharge. There has been considerable discussion about the problem of impotency following resection of the entire aganglionic segment. Some time ago State (1952) suggested anterior resection as the treatment of choice on the assumption that a more extensive operation would result in impotency. We have now operated on six adult males and in each instance ejaculation is normal post-operatively. In view of this, it seems illogical to perform an incomplete resection of the aganglionic segment on the supposition that this will result in impotency. It is very important in doing the resection that the rectum be freed by dissecting right on the bowel wall so that there will be no disturbance of the automatic fibres in the pelvis. This has another useful purpose, for a number of these patients have some defect in bladder function and unless the operation is performed with the greatest care there may be damage enough to produce further atonia of the urinary bladder. In our group of 150 patients there has been one who had some difficulty emptying the bladder post-operatively. She was placed on catheter drainage with a tidal irrigator for two weeks, and during this period bladder tone returned, and her urinary function has remained satisfactory. This complication came early in our experience, and since that time an indwelling catheter is routinely left in place for eight to nine days post-operatively. Stephens, who worked with me in Boston, returned to England, and, using our technique, reported good clinical results (Bodian, Stephens and Ward, 1950). Hiatt, using the same operative technique, reports a large series of good results (Hiatt, 1951).

Early in our experience it became evident that some of our patients with Hirschsprung's disease had urinary bladder dysfunction. This led to studies of bladder function pre-operatively in all of our patients of sufficient age for the performance of satisfactory cystometrograms. A considerable number had some deviation from normal in the cystometrograms. These changes consisted of greater capacity and less vigorous detrusor contractions than normal. In 5% there was considerable derangement of bladder function and dilatation of the upper urinary tract. These patients with megalo-ureters were carefully studied to be sure that they had no mechanical obstruction at the bladder outlet or at the uretero-vesical junction. I began to consider the possible anatomical defects which might explain this association of lesions in the lower colon and in the urinary bladder. The fact that the terminal portion of the colon receives its parasympathetic supply for S-2, 3, and 4 (that is, the pelvic parasympathetic system) suggests that congenital megacolon is a malformation in the pelvic parasympathetic system so that the terminal ganglion cells fail to develop. This would account for the constant location of the lesion in the distal colon. In those patients with the major portion of the colon or all of the colon aganglionic, one would have to postulate a lesion of the terminal portion of the vagal system.

The pelvic parasympathetic system also supplies the detrusor of the urinary bladder, and this fact seems important in the understanding of the patient with megalo-bladder. It would seem reasonable to consider that if one has a congenital lesion of the pelvic parasympathetic system, that this might involve bladder function as well as function of the terminal portion of the colon. Should this be true, it would be logical to postulate three groups of patients: one, patients with a lesion in the colon and an intact urinary bladder; two, patients with a defect in the terminal colon and in the bladder; and third, patients with an intact colon and a defect in bladder function alone. This last group of patients might include some of the patients with idiopathic megalo-ureters.

A group of 35 patients with megalo-ureters has come under my care. Cystometrograms have been utilized in classifying these patients. By our method, a normal cystometrogram begins with an initial zero pressure and gradually rises to 10 or 12 cm. of water pressure before detrusor contractions begin and the bladder empties. In contrast to this, the patients with megalo-bladder have two or three times the capacity of a normal child of comparable age and size, and in addition, the pressure rises to as much as 25 or 35 cm. of water pressure. Detrusor contractions may be absent, or during filling frequent low amplitude contractions may occur, but none during the terminal period of filling. By history these patients void only once or

twice a day and tend to carry large residuals. I have performed ureteral peristalsis studies on these patients, and they have a tracing of active peristalsis that is perhaps somewhat different from normal tracings in that the rate of contraction may be a little slower than that in normal subjects. Ureteral contractions in a normal child occur at 7 to 10-second intervals and the maximum pressure attained by the ureteral contraction varies from 35 to 45 cm. of water. It is obvious that with intravesical pressure almost equalling the pressure generated by a peristaltic wave, the ureter will dilate. It was postulated that these children had defective bladders and that the dilatation of the upper urinary tract was secondary to a malfunctioning bladder. To test this hypothesis, the children were put either on urethral drainage or on suprapubic drainage, and, much to our satisfaction, there was a general improvement in the patients' conditions and in addition, a gratifying decrease in the size of the ureters and hydronephrosis. Prolonged catheter drainage has many complications and we have resorted to a different form of treatment in the last few years. In order to reduce the resistance to voiding, extensive resections of the bladder neck were performed, and by instructing the children to void frequently and to make a great effort to empty the bladder completely, gratifying results have been achieved. The rationale of this is to have these patients use the bladder where the range of pressure is near normal. An equally important part of the management of these children is to combat urinary infection. The parents and the local physicians are informed that these children must be considered in the same category as patients with rheumatic heart disease. They must be carefully observed for a number of years. They should see their physician at least once a month for residual determinations and urine analysis. A wide variety of antibiotics is used to control the infection. It is most gratifying to see how much can be accomplished with these children providing the child, the family and the local physician are willing to persevere (Fig. 3). The precise nature of the bladder dysfunction is difficult to ascertain. An opportunity presented itself to study eight bladders removed from patients who had succumbed to the disease. Fortunately, the radiographs and clinical records were available for study. The specimens removed at necropsy included the kidney, ureters, bladder and urethra. There was no evidence of mechanical obstruction in the lower urinary tract. The entire bladder was cut into blocks and imbedded. It is important to emphasize that the whole organ was thus imbedded,

FIG. 3A.—Cystogram of patient with megalo-bladder and megalo-ureters due to an aparasympathetic bladder. Note bilateral reflux. Residual 150 ml.

FIG. 3B.—Cystogram on same patient three months after resection of the bladder neck. Residual 15 ml. This patient had adhered to a programme of voiding five times a day and once during the night.

and histological sections were studied from each paraffin block. Therefore, the whole bladder was studied. We took a similar number of normal bladders and studied them in the same manner in order to determine the number and distribution of ganglion cells. It was found that in the normal specimen the concentration of ganglion cells was greatest in the peri-ureteral area on each side and that the concentration diminished rapidly in all directions from the ureteral orifice (Swenson, MacMahon, Jaques and Campbell, 1952).

In studying the specimens from patients with megalo-ureters and megalo-bladder a striking diminution in the number of ganglion cells was discovered. In some of the specimens the ganglion cells were extremely difficult to find. This lends support to our conception that these patients have a parasympathetic defect in the urinary bladder, and because of abnormal innervation they have enlarged bladder, high intravesical pressure, difficulty in emptying, and carry large residuals. A second group of patients with megalo-ureters was encountered which does not fit into this category. This group of patients has an essentially normal bladder, but upon study of ureteral function peristalsis is found to be absent. In one patient there was a rhythmic activity in the ureter, but actually the tracing was the reverse of that seen in normal patients so that the ureters seem to be intermittently dilated rather than contracted (Fig. 4). The cause of this dysfunction is obscure. Infection may play a role, but it is not the major factor, for a number of patients with infection have been demonstrated to have normal peristalsis. A congenital malformation is probably the underlying pathology. This is difficult to establish histologically for there are few ganglion cells in a normal ureter so that study of these neural elements has not proved helpful. Massive chronic dilatation probably plays a role. However, normal peristalsis has been demonstrated in ureters many times normal in size.

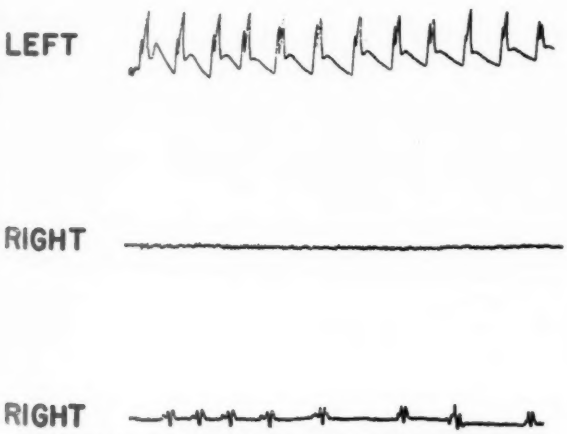

FIG. 4.—Ureteral peristalsis tracings from a patient with a normal bladder and right megalo-ureter. Note normal peristalsis in left ureter. On the right side no peristalsis could be detected during two long periods of observation.

One practical point should be stressed and that is in the group with aparasympathetic bladders; nothing is gained by operation on the lower end of the ureter; in fact, irreparable damage may be done. I have seen two patients who, after bilateral resection of the lower ends of the ureter and reimplantation, were unable to void. These patients probably had a congenital defect in the pelvic parasympathetic system and the operative procedure destroyed what remaining innervation they had.

This group of patients has been the most difficult to treat. They have done poorly as a group and all that one can do is control the infection and, as their retention of nitrogenous products mounts, one can only resort to nephrostomy.

After careful study our group of 35 patients fall into the following groups: 17 had aparasympathetic bladder and four of these had Hirschsprung's disease as well; nine had absent ureteral peristalsis; four had cord bladder; three had ureterocoeles; and two had agenesis of the abdominal wall.

Summary

Hirschsprung's disease is due to a congenital malformation in the pelvic parasympathetic system. The absence of ganglion cells in the distal colon accounts for the absence of peristalsis in this segment of the intestinal tract. There is one group of patients with megalo-ureters and megalo-bladder in which the pathology is a defect in the parasympathetic innervation in the bladder. Ureteral peristalsis is essentially normal in this group of patients with megalo-ureters and megalo-bladder. There is a second group of patients with megalo-ureters, either unilateral or bilateral, in which the bladder is normal and peristalsis is absent or highly abnormal.

REFERENCES

Bodian, M., Stephens, F. D. and Ward, B. C. H. (1950). *Lancet*, **1**, 19.
Dalla Valle, A. (1920). *Pediatria, Napoli*, **28**, 740.
Hiatt, R. B. (1951). *Ann. Surg.*, **133**, 321.
Robertson, H. E. and Kernohan, J. W. (1938). *Proc. Mayo Clin.*, **13**, 123.
State, D. (1952). *Surg. Gynec. Obstet.*, **95**, 201.
Swenson, O. and Bill, A. H., Jr. (1948). *Surgery*, **24**, 212.
——, MacMahon, H. E., Jaques, W. E. and Campbell, J. S. (1952). *New Engl. J. Med.*, **246**, 41.
——, Rheinlander, H. F. and Diamond, I. (1949). *Ibid.*, **241**, 551.
Tiffin, M. E., Chandler, L. R. and Faber, H. K. (1940). *Amer. J. Dis. Child.*, **59**, 1071.
Zuelzer, W. W. and Wilson, J. L. (1948). *Ibid.*, **75**, 40.

HIRSCHSPRUNG'S DISEASE IN THE NEONATAL PERIOD*

BY

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I do not intend to give an account of the changing philosophies concerning the aetiology and treatment of the disease which followed upon Hirschsprung's (1888) first presentation of it as an entity in 1886. It required about 60 years of vague or wrong ideas to find the key to our present knowledge. That happened when our attention was diverted from the characteristic megacolon and was focused on a less conspicuous and apparently normal distal part of the bowel.

In 1946 a study of the early stages of the disease in 10 newborn infants (Ehrenpreis, 1946) revealed that the megacolon was not present at birth but developed only secondarily. Hirschsprung's (1888) assumption that constipation was caused by congenital megacolon could thus be reversed: the megacolon proved to be caused by the congenital constipation.

During the next few years definite proof of the cause of congenital constipation was presented. Evidence was produced that a narrow or normal-looking segment of rectosigmoid distal to the dilated colon was the primary pathology in Hirschsprung's disease (Swenson and Bill, 1948). Neurohistological investigations demonstrated that the entire distal segment lacked ganglion cells (Whitehouse and Kernohan, 1948; Bodian, Stephens and Ward, 1949; Swenson, Rheinlander and Diamond, 1949). Removal of the distal segment cured the disease (Swenson and Bill, 1948).

Finally, a most important advance was made by the subdivision of the group of idiopathic megacolon into two different disease entities (Bodian *et al.*, 1949). Only about half of the cases of idiopathic megacolon showed the clinical, radiological and neurohistological signs which had been demonstrated as typical of Hirschsprung's disease. The other half disclosed a different pattern of symptoms and signs, the main diagnostic difference being the absence of a distal aganglionic segment (Figs. 1-2). This division has made it possible to account for the controversial results of earlier investigations and, still more important, to select the cases suitable for surgical treatment by rectosigmoidectomy.

Ehrenpreis's (1946) study of the onset and early development of the disease in 10 newborn infants disclosed a disease pattern which is essentially different from the typical and well-known picture of chronic constipation, abdominal enlargement and radiological megacolon in later infancy and childhood.

Illustrative Case

In 1943 a 3-day-old infant was brought to the Crown Princess Louisa's Hospital in Stockholm with a tentative diagnosis of intestinal obstruction. Only a small amount of meconium had been passed. On the third day of life the baby started to vomit and the abdomen became increasingly distended. Clinical examination revealed nothing abnormal except the abdominal distension. A straight radiograph showed a moderate and diffuse gaseous distension of the small intestine and of the colon, so that any obstruction would be situated in the distal part of the colon. A barium enema, however, failed to reveal any obstruction, nor was any other abnormality demonstrable in the large bowel. The diagnosis was difficult. Clinically, the baby presented all the symptoms and signs of an intestinal obstruction. There seemed to be no reason why this otherwise healthy baby should develop a paralytic ileus. A diagnosis of Hirschsprung's disease was considered, and it was suggested that the typical dilatation of the colon might not necessarily be a congenital lesion but might have developed only as a result of congenital constipation. On reviewing the literature, however, no substantial support for such an assumption could be found, and the course of the case at first also seemed to refute the diagnosis. On evacuation of the barium enema, the bowels began to move, the abdominal distension subsided, and vomiting ceased. During one week of subsequent observation in the ward the baby appeared and behaved like an entirely normal and healthy infant. There were no feeding difficulties, no vomiting, no abdominal distension and normal bowel movements. At the age of 10 days the baby was discharged with a tentative diagnosis of meconium constipation. Not many days later, however, signs of intestinal obstruction again developed. At the age of 14 days the baby was readmitted with vomiting and abdominal distension.

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

FIG. 1.—Narrow segment in Hirschsprung's disease.

No stools and no gas had been passed for 24 hours. A barium enema still failed to support a diagnosis of Hirschsprung's disease and evacuation of the enema again started bowel movements. After this, obstructive attacks reappeared with increasing frequency and intervals of apparent health successively came to be shorter and shorter. At the age of 5 weeks the classical disease picture of chronic constipation with abdominal enlargement developed, and the diagnosis of Hirschsprung's disease could also be confirmed radiologically.

This case was a very interesting experience. It exhibited a new picture of the disease, which in essential features diverged from the classical description. It now seemed desirable to find out if this picture was typical of the neonatal period or if it was only an exception.

The opportunity for further studies was soon offered. Three additional cases were admitted to the hospital within a short time and about a year later a fifth case appeared. From our files of Hirschsprung's disease five more cases could be traced back to the newborn period both clinically and radiologically. All of these 10 cases showed the same clinical pattern of repeated attacks of intestinal obstruction with intervals of apparent health and radiological absence of the typical megacolon during the neonatal period.

The three main obstructive symptoms of constipation, abdominal distension and vomiting have been tabulated according to their occurrence and day of onset in this series (Table 1). All of the

TABLE 1

CLINICAL ONSET OF MEGACOLON IN 10 NEWBORN INFANTS

Case No.	1	2	3	4	5	6	7	8	9	10
Constipation ..	1	1	1	1	1	1	1	?	?	1
Distension ..	3	3	4	2	2	—	5	2–3	2	3
Vomiting ..	3	—	2	—	1	1	5	2–3	7	3

The numbers define the age in days at appearance of the respective symptoms.

obstructive symptoms appeared fairly constantly during the first few days of life.

The radiological findings during the first week of life were evaluated on the basis of barium enema studies of 100 normal newborn infants. No significant dilatation of the colon was demonstrable at examination in the Hirschsprung series. As an example, three of the Hirschsprung colons have been photographed together with three normal colons (Fig. 3). I doubt if even the most experienced among my listeners would be able to distinguish between them.

In all the cases the obstructive symptoms subsided after the barium enema. The subsequent

FIG. 2.—Rectal dilatation in chronic constipation.

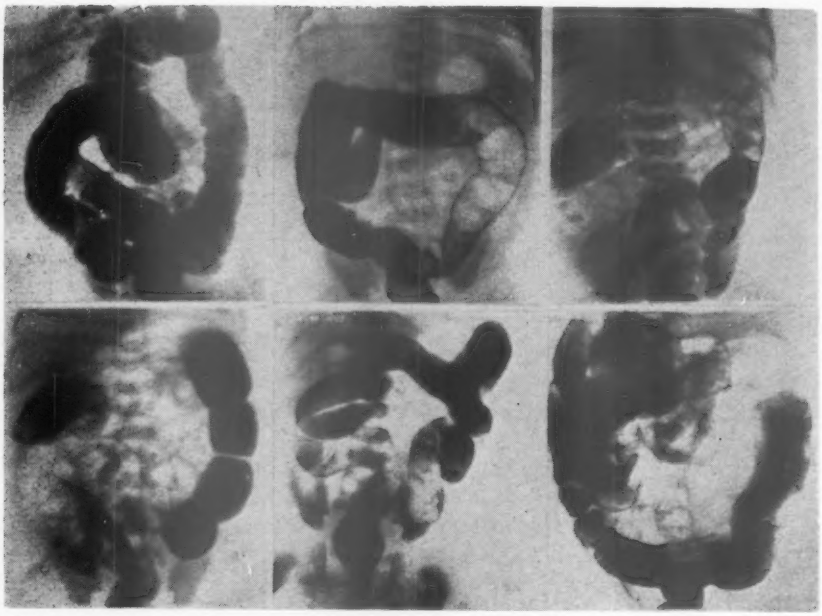

FIG. 3.—Radiographs of colon in three newborn infants with Hirschsprung's disease and three normal newborn infants.

course of the disease appears in Table 2. In the majority of cases, a period of apparent health followed upon the relief of the initial obstruction. The length of this symptom-free period varied between one week and two and a half months in this series. Subsequently, obstructive periods alternated with intervals of apparent health for varying periods of time until the final transition into the classical morbid picture. The three main clinical characteristics of this picture, compared with the early stages, are chronic constipation, persistent abdominal enlargement, and absence of vomiting. As appears from Table 2, all these changes occurred more or less simultaneously in every single case, at ages varying between 3 or 4 weeks and 8 or 10 months.

Radiologically, all the cases developed the typical picture of megacolon. In four, barium enemas were repeated frequently enough to establish with a fair degree of accuracy the time required for the development of the radiological megacolon picture. The shortest time was 18 days and the longest time three and a half months (Fig. 4).

A synopsis of clinical and radiological data concerning the development of the disease picture suggests a pathophysiological explanation for the differences between the early and late stages. As long as the colon is of more or less normal size, the obstruction has an almost unlimited retrogressive effect, and the picture of acute intestinal obstruction results. Intervals of apparent health with normal bowel movements might be explained by reserve capacities of the still unaltered proximal colon, at least in cases with a short aganglionic segment. With the development of colon dilatation a kind of elastic reservoir is provided, which absorbs most of the retrogressive tendency. At this stage the proximal colon becomes more and more insufficient and finally loses power to overcome the obstruction. Thus the picture of chronic constipation with abdominal enlargement results.

What still remains to be explained is the occurrence of free intervals in cases with a long aganglionic segment. I recently saw a case with an extremely long aganglionic segment, comprising the whole colon except the caecum. This child had developed a transitory intestinal obstruction during the first few days of life, which soon subsided. After this he appeared perfectly normal with daily bowel movements for about four months, when intestinal obstruction again developed and failed to respond to conservative treatment. A caecostomy was performed and biopsies were made from the sigmoid, transverse and proximal ascending colon. Microscopic examination revealed absence of ganglion cells from all three of these specimens. Since caecostomy the patient has thrived and is now

TABLE 2
DEVELOPMENT OF DEFINITE (CLASSICAL) CLINICAL PICTURE

Case No.	1	2	3	4	5	6	7	8	9	10
Period of apparent health following onset	1 w.	2 w.	1 w.	—	2½ m.	—	4 w.	—	1 w.	1 w.
Beginning of persistent constipation ..	—	8 m.	3 w.	birth	4½ m.	birth	4 m.	birth	4 w.	3 w.
Beginning of persistent abdominal enlargement	1 m.	8 m.	3 w.	?	<5½ m.	>4 m.	<10 m.	?	?	—
Ceasing of vomiting	1 m.	10 m.	5 w.	—	6 m.	6 m.	8 m.	<1 y.	6 w.	6 w.

awaiting colectomy with anastomosis between the lower rectum and the detached caecostomy or terminal ileum.* I have nothing to offer in the way of explanation for the long period of normal bowel movements in a case like this.

This specific disease pattern of the neonatal period and early infancy has been confirmed by several investigators (Bodian *et al.*, 1949; Burnard, 1950; Swenson, 1951; Potts, Boggs and White, 1952).

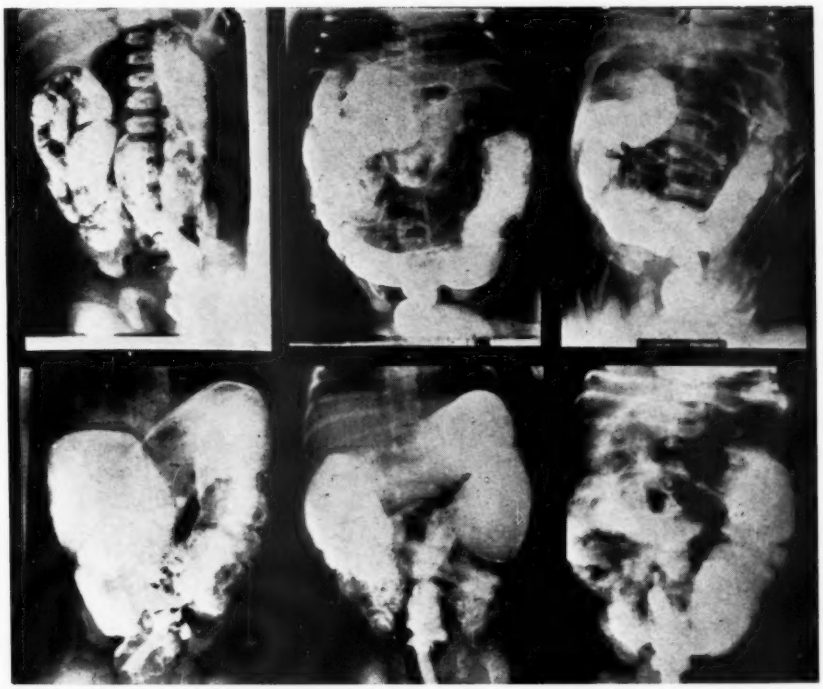

FIG. 4.—Radiological development of megacolon in three cases. Upper row: Newborn period. Lower row: Same cases at 3½ months, 5 weeks, and 6 weeks, respectively.

Diagnosis

Before acquaintance with the early disease picture the diagnosis used to be something of a problem. Many cases were operated upon in the neonatal period with a diagnosis of intestinal obstruction. Usually operation was of no great help either as a diagnostic or a therapeutic measure, as no cause for the intestinal obstruction could be demonstrated. Now the diagnosis should offer no real difficulties. Every newborn baby presenting with signs of a low intestinal obstruction should be submitted to radiological examination. First the diagnosis of intestional obstruction has to be confirmed by plain films, showing distension of intestinal loops with fluid levels. Then, the cause for the obstruction has to be determined by barium enema studies. If a distal small but patent part of the colon (microcolon) fills, and if filling of the remainder of the colon proves impossible, there is probably an atresia in the colon. If the whole of the colon shows the characteristic microcolon (Fig. 5), there is a total obstruction above the colon. This obstruction might be due either to an atresia within the small bowel or to a meconium ileus. A more or less normal-looking or slightly distended colon (Fig. 3) suggests an obstruction within the distal part of colon. There are two possible causes for such an obstruction. The more common cause is Hirschsprung's disease, more rarely simple meconium constipation. The immediate differentiation between these two conditions is often impossible, which, however, is of no great practical importance. In either event the obstructive symptoms usually subside after a barium enema. Recurrence of obstructive symptoms suggests the diagnosis of Hirschsprung's disease, and repeated barium enemas will soon confirm this diagnosis (Fig. 4).

These simple diagnostic considerations were adopted about 10 years ago in Stockholm, and they have proved to be very helpful. Admittedly, there are other rare causes for a low intestinal obstruction in the newborn. Provided that due attention is paid to the clinical picture as a whole as well as to the radiological findings, the danger of missing a diagnosis of intestinal obstruction requiring surgery is slight.

The outlook for a newborn baby with Hirschsprung's disease used to be extremely poor. Out of the 10 patients discussed above, five died, four of them before the age of 1½ years. Similar and even higher mortality rates are given in other comparable series (Burnard, 1950; Potts *et al.*, 1952; Hirschsprung, 1904; Ask-Upmark, 1930). With our present concepts of the aetiology, diagnosis and treatment of Hirschsprung's disease a definite change to a better prognosis has been accomplished for all age groups, even though the neonatal period is still a critical one. Larger series of newborn babies treated according to modern principles are still

* Since this paper was submitted for publication a total colectomy with anastomosis between the terminal ileum and lower rectum has been successfully performed in this case.

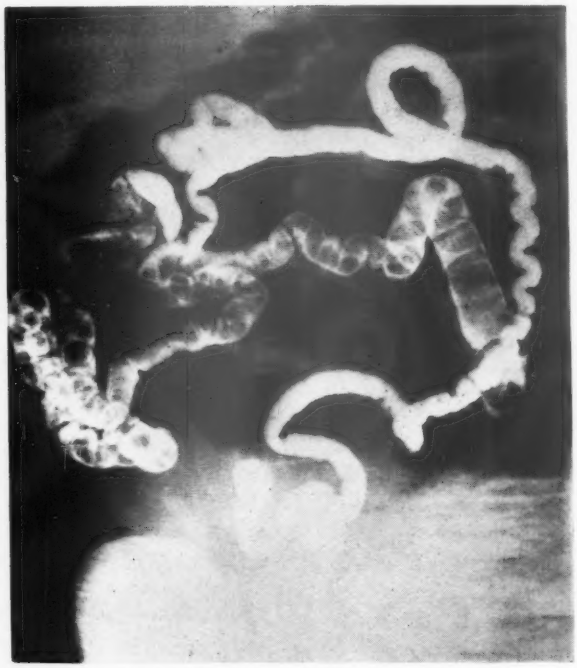

FIG. 5.—Microcolon and terminal ileum.

lacking, so I cannot give any exact figures as to present mortality rates of the initial stages of the disease.

Treatment

Present opinions as to the principles of treatment are practically unanimous. Resection of the aganglionic distal segment with anastomosis between the proximal dilated bowel and the lower rectum is the only treatment which eradicates the cause of the disease. This operation has been generally adopted as the treatment of choice and should be performed in every case with a definite diagnosis of Hirschsprung's disease.

When dealing specifically with the early stages of the disease the following considerations have to be taken into account. As long as the disease presents as attacks of recurring intestinal obstruction with intervals of apparent health there is generally no need for any kind of treatment, except occasional enemas or colonic irrigations. During this stage delimitation of the distal narrow segment is usually impossible or uncertain, either by radiological or surgical exploration. Should an obstructive attack fail to respond to conservative measures an emergency arises, which has to be dealt with by colostomy decompression of the proximal bowel. During an obstructive attack delimitation of the aganglionic segment might be possible, but resection in this stage would still not be the proper procedure because of the ileus. Resection is thus as a rule not practicable in early infancy.

As soon as the disease has entered into the fully developed stage, operation should be performed, usually as a one-stage procedure. Only if abdominal distension is not relieved by colonic irrigations, or when the patient's general condition does not seem to warrant the major procedure of rectosigmoidectomy, a preliminary colostomy has to be performed. The results of this operation have been most gratifying. Up to now reports of about 300 cases treated by rectosigmoidectomy have been published (Swenson, 1954; Bodian, 1952; Grob, 1952; Hallenbeck and Waugh, 1952; Ehrenpreis, 1952; Hiatt, 1951). The overall mortality rate is about 5%. Except for some few failures due to incomplete resections, all of the patients have been relieved of their symptoms. Some of the patients have been followed now for six years, and so far, no recurrences have been reported after radical resection. Post-operative complications have been few and transitory.

These excellent results testify to the physiological soundness of the present concept of Hirschsprung's disease and mark the rectosigmoidectomy operation as the first and only rational treatment.

REFERENCES

Ask-Upmark, E. (1930). *Bruns' Beitr. klin. Chir.*, **151**, 73.
Bodian, M. (1952). *Practitioner*, **169**, 517.
——, Stephens, F. D. and Ward, B. C. H. (1949). *Lancet*, **1**, 6.
Burnard, E. D. (1950). *Brit. med. J.*, **1**, 151.
Ehrenpreis, T. (1946). *Acta chir. scand.*, **94**, Suppl. 112.
—— (1952). *Ibid.*, **102**, 251.
Grob, M. (1952). *Helv. chir. Acta*, **19**, 336.
Hallenbeck, G. A. and Waugh, J. M. (1952). *Surg. Clin. N. Amer.*, **32**, 1203.
Hiatt, R. B. (1951). *Ann. Surg.*, **133**, 321.
Hirschsprung, H. (1888). *Jb. Kinderheilk*, **27**, 1.
—— (1904). In Grancher, J. and Comby, J. *Traité des maladies de l'enfance*, 2nd ed., vol. 2, p. 241. Paris.
Potts, W. J., Boggs, J. D. and White, H. (1952). *Pediatrics*, **10**, 253.
Swenson, O. (1951). *Ibid.*, **8**, 542.
—— (1954). *J. Amer. med. Ass.*, **154**, 651.
—— and Bill, A. H. (1948). *Surgery*, **24**, 212.
——, Rheinlander, H. F. and Diamond, I. (1949). *New Engl. J. Med.*, **241**, 551.
Whitehouse, F. R. and Kernohan, J. W. (1948). *Arch. intern. Med.*, **82**, 75.

INTESTINAL OBSTRUCTION IN THE NEWBORN*

BY

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There are many causes of intestinal obstruction in the newborn baby. Individually none of them are common conditions and yet together they make up a group of some importance. Rickham (1952) has estimated that up to 950 babies are born in Great Britain each year with congenital defects amenable to surgery and the obstructions form a large part of these.

This subject has been well discussed in several recent articles (Gross, 1953; Jolleys, 1952; Santulli, 1954; Swain and France, 1954, etc.). In this paper I wish to refer to particular aspects which I have had the opportunity of observing during the past five years, mainly at The Hospital for Sick Children, Great Ormond Street.

Diagnosis

Delay in diagnosis is still an important factor in failure to relieve these obstructions. For example, of 21 recent cases of jejuno-ileal atresia admitted to this hospital, 18 babies began vomiting on the first day of life and yet only 17 were admitted by the third day. Why should this two-day lag period still exist?

The effects of obstruction are the same as at any other age. Absolute constipation occurs but the early passage of meconium may be apparently normal while the bowel below the obstruction is emptying. There is distension, but in the early stages it is difficult to assess in the naturally protuberant abdomen of the newborn, and in duodenal obstructions may be entirely lacking in a baby who vomits easily. Vomiting remains as the most reliable early sign in the majority of cases. There are many other reasons for neonatal vomiting and this seems to be the reason why clinicians may be slow to suspect obstruction. However, bile-stained vomit such as one sees in cases of obstruction below the bile ducts is not frequent.

Two large maternity hospitals were able to discover only two cases of true bile-stained vomiting occurring in the absence of an organic cause in mature babies during a year's observation during which about 4,000 babies were delivered. In a small series of 106 babies at Queen Charlotte's Maternity Hospital, London, every vomit was charted on a special form so as to ensure that small vomits were not passing unrecorded because the nursing staff considered them trivial. As would be expected a large proportion of the babies produced some vomitus but none was green (Table 1). Eight produced yellow vomitus which has often been attributed to bile staining. Four specimens of vomit were examined by Dr. G. H. Lathe. Bile pigments (Fouchet's reagent) were absent and the spectral absorption curve of extracts of vomitus indicated that the main pigments were of a carotenoid type known to be present in large amounts in colostrum (Dann, 1936). This may explain the misconception that bile-stained vomitus is not uncommon in normal newborn babies.

TABLE 1

VOMITING IN FIRST DAYS OF LIFE

No. of Births	No. Vomiting	Colour of Vomit			
		White	Yellow	Brown	Green
106*	23	12	8	3	0

* Vomiting persisted over 24 hours in eight. There were no relevant abnormalities in the babies.

A second factor encouraging delay is the apparent well-being of the obstructed baby in the early stages. The newborn baby is prepared to live on his resources for some days and does not begin to look ill until secondary vascular changes develop in the distended or volved bowel, or until vomiting has been prolonged for some days. Thus there is a reluctance to transfer a fit looking baby to a surgical unit.

Fortunately, however, the most valuable confirmatory investigation is one which can be carried out in any hospital and with negligible disturbance to the patient. Plain radiographs of the abdomen are usually sufficient to clinch the diagnosis if properly interpreted. The basic feature in most cases is the presence of fluid levels in progressively distending

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

loops of bowel. When the presence of obstruction is confirmed, laparotomy should follow. Delay for further study can be dangerous and the exact nature of the lesions will be better diagnosed by direct observation.

Barium meals are unnecessary and may be dangerous, except perhaps, in suspected incomplete obstructions. A small barium enema may prove to be of value in Hirschsprung's disease. I have not found Farber's test helpful. It is only positive in complete atresias which are clearly diagnosable by clinical examination and plain radiographs, and when negative only excludes this one cause of obstruction.

One must be on guard for other organic causes of neonatal vomiting of which the chief are birth injuries of the brain and infections. Cretinism can also cause constipation to the extent of obstructive vomiting, and cystic fibrosis of the pancreas may cause constipation and vomiting without necessarily progressing to the full picture of obstruction by meconium ileus.

Persistent vomiting may also occur in oesophageal hiatus hernia at this age.

Malrotation

These conditions have been admirably described by Frazer and Robbins (1915) and Dott (1923), and Ladd's (1932, 1933) procedure of unrotating the gut usually relieves the obstruction equally well.

When volvulus has been present for some time before birth the bowel has grown in its twisted position and its unwinding may be more difficult and will leave raw areas without complete peritoneal covering. Fig. 1 shows a peritoneal band found extending from the midline to the beginning of the jejunum. I think it is important after dividing the transduodenal bands of Ladd to continue clearing the first part of the jejunum lest obstruction at this point persist. I imagine this band to be an attempt at the formation of a ligament of Treitz in the presence of a misplaced duodenal flexure (Spencer, 1951). The ileocolic adhesion also shown is not uncommon: it may form a pocket between bowel and mesentery. One of our cases died of strangulation of a loop of ileum herniated through such an orifice. Results have been good, 12 of 15 cases in the past five years surviving, and the three deaths all in complicated cases.

Duodenal Atresia and Stenosis

Those duodenal atresias situated above the ampulla of Vater are likely to be diagnosed later as the vomit consists only of gastric contents and does not become bile stained. However, the very fact that the biliary and intestinal secretions are not lost and the fact that the lesion cannot lead to strangulation means that delay is here less serious than in atresia lower down.

More disturbing is the appallingly high incidence of mongolism in this series, 10 of a consecutive 32 cases studied by Bodian, White, Carter and Louw (1952). The condition can be recognized at birth by careful inspection of the facial, cranial, and manual stigmata. Treatment may then become a matter for ethical consideration.

Louw (1952) showed that in the earlier cases at Great Ormond Street there was a high mortality after gastro- and duodeno-jejunostomy and that this was not usually due to technical failure in the operation. It was the result of failure to manage the difficult 10 days until the anastomosis would transmit an adequate amount of milk to sustain the baby. Ehrenpreis and Sandblom (1949) greatly improved their results by the use of a transanastomotic tube to allow of early feeding. I have no personal experience of this and perhaps improved parenteral therapy should render it unnecessary, but the idea is attractive and our results still leave much to be desired—only five survivals out of 18 cases during the past five years.

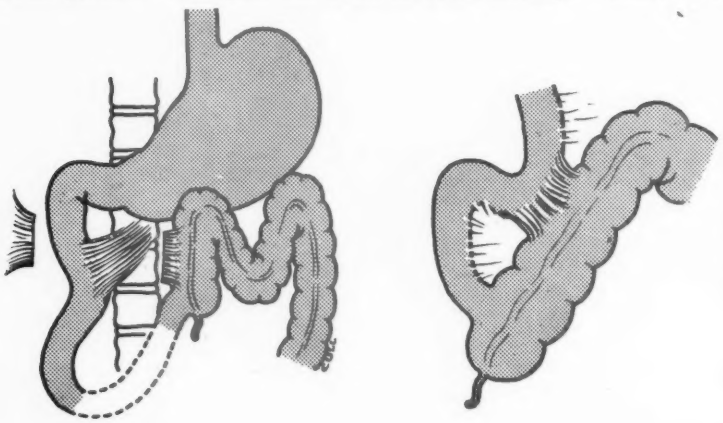

FIG. 1.—Drawing to show the binding down of the beginning of the jejunum and ileo-colic adhesion in a case of malrotation.

Jejuno-ileal Atresia and Stenosis

Diagnosis is here straightforward but more urgent because delay causes vascular changes in the bowel wall by increased distension and eventual perforation. The results of simple anastomosis have been poor in our hands. Louw (1952) showed that in cases

treated reasonably early the main cause of death, unlike the duodenal cases, lay in the failure of the anastomosis to function.

Hoppu (1951) suggested that any primary anastomosis would inevitably be unreliable in bowel of this small calibre and that a prosthesis would be necessary to prevent plugging and functional obstruction. In experiments with baby rabbits of a comparable bowel size and structure he had a mortality of 58·3% from obstruction with end-to-end anastomosis. I repeated these experiments and found my mortality to be only 30% (Fig. 2) without the assistance of parenteral fluids or chemotherapy. The use of other types of anastomosis suggested that reasonable success could be obtained and that the results depended on the technical convenience of the method in the particular animal used rather than on the size of the aperture created.

I believe that the cause of failure in direct anastomoses in cases of atresia lies in the dilated and hypertrophied proximal bowel and not in the unexpanded distal bowel. The obstruction is an acute one superimposed on a chronic foetal one and as a result several inches of bowel proximal to the obstruction are not only dilated but also anatomically enlarged and hypertrophied. This gut contracts actively but is nevertheless ineffective in propulsion. It does not seem to close the lumen sufficiently to have a directional effect on fluid contents.

The first two cases of jejuno-ileal atresias on which I operated were both diagnosed so late as to necessitate resection of the proximal bulb of bowel for

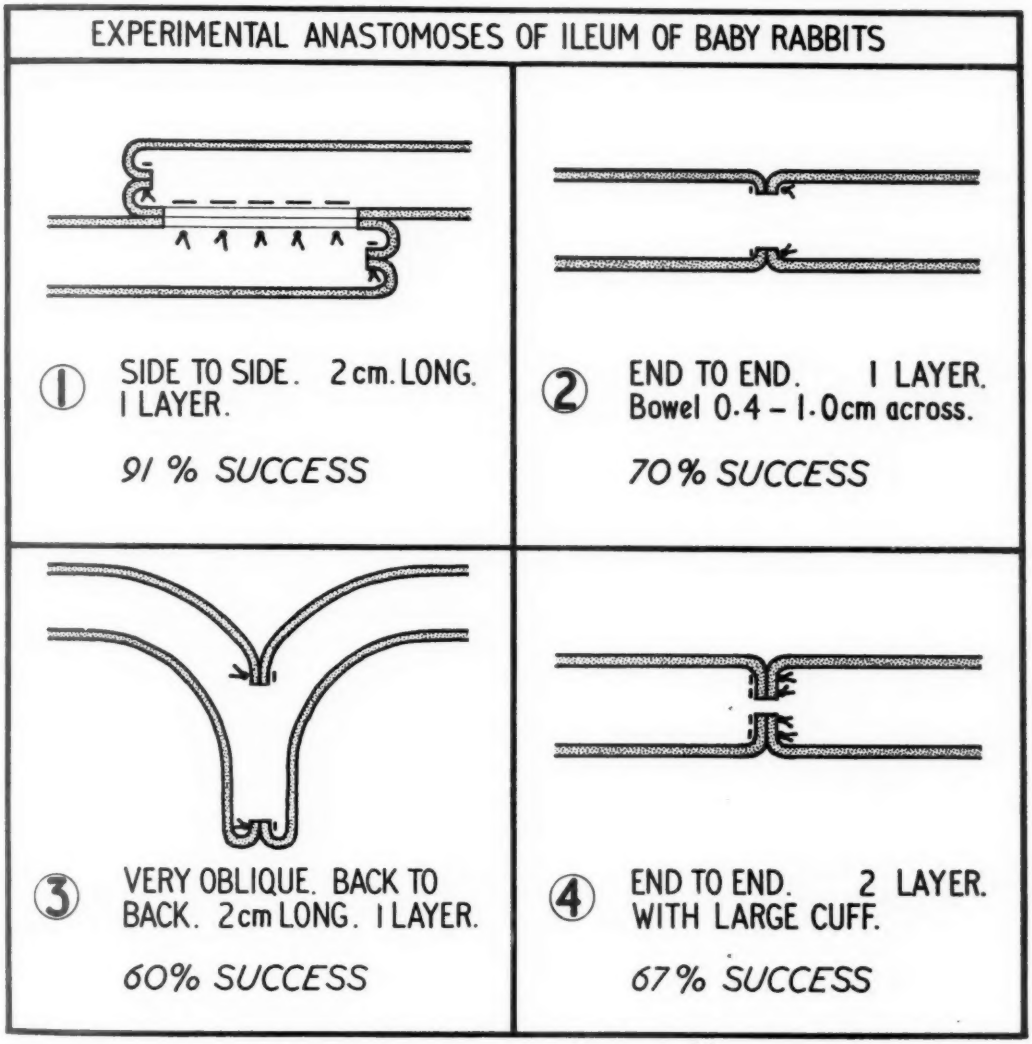
EXPERIMENTAL ANASTOMOSES OF ILEUM OF BABY RABBITS
1
SIDE TO SIDE. 2 cm. LONG. 1 LAYER.
91 % SUCCESS
2
END TO END. 1 LAYER. Bowel 0·4 – 1·0 cm across.
70 % SUCCESS
3
VERY OBLIQUE. BACK TO BACK. 2 cm LONG. 1 LAYER.
60% SUCCESS
4
END TO END. 2 LAYER. WITH LARGE CUFF.
67 % SUCCESS

FIG. 2.—Results of experimental anastomoses of ileum in 70 young rabbits.

incipient gangrene; both survived. The third, a case of high jejunal atresia, was diagnosed early and in excellent health and I performed a simple end-to-oblique anastomosis. All went well until the feeds were increased to full requirements when vomiting recurred. Several trials showed that the anastomosis would only pass about half the food required. A second laparotomy was performed at which the anastomosis was found to be patent although it was not functioning. A long lateral anastomosis was made around it. Nevertheless, the same trouble was experienced after operation and the baby died aged 5 weeks with a widely patent anastomosis, full expansion of the distal bowel, and yet persistence of the distension of the bowel proximal to the anastomosis.

During the baby's life the ward sister pointed out to me that during feeds the milk could be felt to enter the dilated bulb rapidly but that it stayed churning there until part was returned by vomiting. There was no paralytic ileus but the bulb was ineffective in propulsion.

Animal experiments are in progress to study this phenomenon. Fig. 3 shows the principle underlying the apparatus which I have called a 'gut pump'. The outlet is raised 1 cm. above the inlet so that the gut has to do measurable work to transmit the fluid. It was noted that the regular longitudinal contractions recorded in the usual isolated organ bath preparations had no propulsive value but that the intermittent stripping contractions in which the segment contracted down hard from end to end produced a sudden passage of fluid. Normal bowel worked best with an inlet pressure of about 2·5 cm.

The next stage was to make a hypertrophied loop

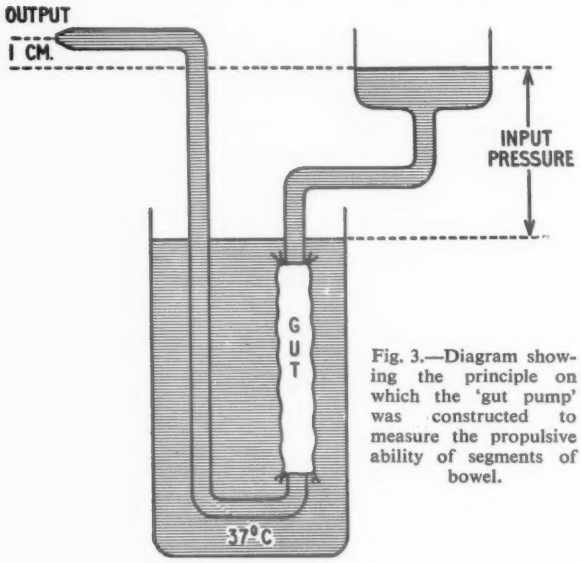

Fig. 3.—Diagram showing the principle on which the 'gut pump' was constructed to measure the propulsive ability of segments of bowel.

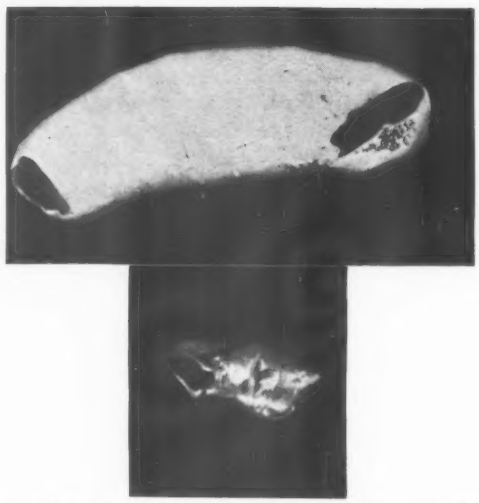

FIG. 4.—Hypertrophied segment of rabbit ileum compared with a normal segment.

of bowel by reversing a segment of ileum, restoring continuity by two end-to-end anastomoses and sacrificing the animal a few weeks later. The bowel above the reversed segment enlarged and hypertrophied as shown in Fig. 4. In the organ bath normal bowel worked best at 2·5 cm. inlet pressure transmitting 45 ml. in 10 minutes. The hypertrophied segment, although moving vigorously, was quite inefficient at this pressure, transmitting only 4 ml. in 10 minutes. If the inlet pressure was raised to 6 cm. then the normal bowel was overstretched and failed, whereas the hypertrophied segment became most efficient, sending over 106 ml. in 10 minutes. In other words, the hypertrophied segment has adjusted itself to abnormal obstructive conditions and is inefficient when a normal intestinal gradient is restored.

TABLE 2

JEJUNO-ILEAL ATRESIA, 1949-54

No. with Complete Atresia	No. with Stenosis	Results of Treatment* by	
		Simple Anastomosis	Resection and Anastomosis
27	6	15 operations, 5 survivals (33%)	15 operations, 10 survivals (67%)

* Deaths included five early post-operative deaths and one 'biochemical' death.

Three cases were not operated upon: two similar cases associated with meconium ileus, volvulus and meconium peritonitis which died during attempted resuscitation, and one case which died soon after operation on an associated oesophageal atresia. This infant also was premature, had kernikterus and other abnormalities.

These clinical and experimental observations led me and some of the surgeons at the hospital to undertake routine resection of the hypertrophied segment of bowel before anastomosis. Other surgeons per-

sisted with simple anastomosis and relied on careful parenteral fluid therapy to keep the baby alive until the bowel function was re-established. The results are shown in Table 2, two-thirds of the cases surviving after resection and only one-third without it.

The technique is illustrated in Fig. 5. From 15 to 25 cm. of bowel are resected and the proximal bowel sucked clear of content which may otherwise tend to inspissate and block the anastomosis. The distal bowel is inflated with saline to clear any concretions and to exclude any further atresias. Anastomosis is then performed with one layer of interrupted silk mattress sutures. The proximal bowel is cut sufficiently obliquely to make the opening 2 cm. long. The distal bowel is cut along its antimesenteric border for a like distance. The bowel is controlled by stay sutures and no clamps are used.

Aetiology

I have studied all our cases of jejuno-ileal atresia whether or not there were associated and more impressive anomalies such as meconium peritonitis. In this way I think that a progression of cases can be shown from simple complete atresia with a blind bowel end to cases with only a slight lesion of the bowel wall, and that these throw some light on the aetiology. Thirty-three cases of jejuno-ileal atresia have been admitted during the past five years.

The commonly quoted view of their origin is that

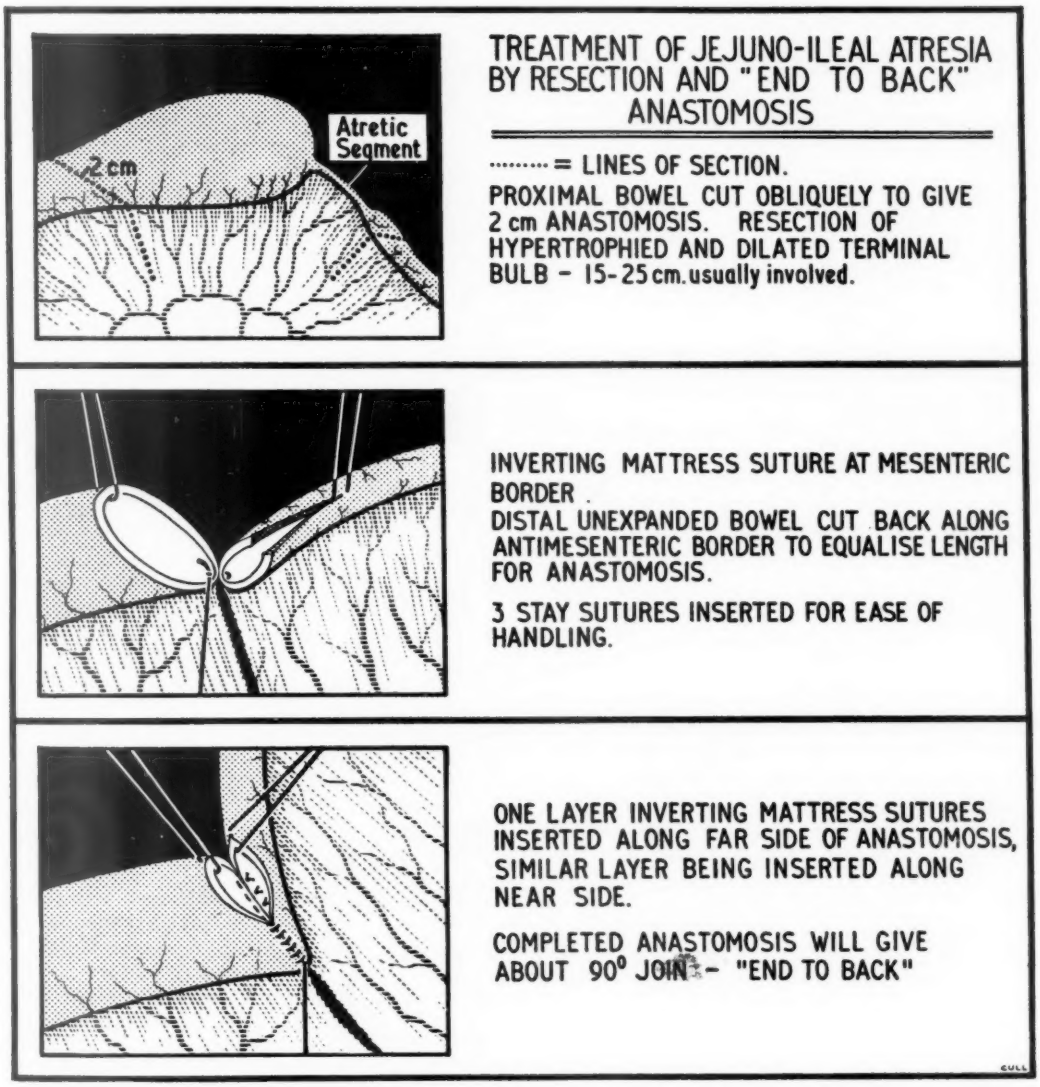
Atretic Segment
2 cm
TREATMENT OF JEJUNO-ILEAL ATRESIA BY RESECTION AND "END TO BACK" ANASTOMOSIS
..... = LINES OF SECTION.
PROXIMAL BOWEL CUT OBLIQUELY TO GIVE 2 cm ANASTOMOSIS. RESECTION OF HYPERTROPHIED AND DILATED TERMINAL BULB – 15-25 cm. usually involved.
INVERTING MATTRESS SUTURE AT MESENTERIC BORDER.
DISTAL UNEXPANDED BOWEL CUT BACK ALONG ANTIMESENTERIC BORDER TO EQUALISE LENGTH FOR ANASTOMOSIS.
3 STAY SUTURES INSERTED FOR EASE OF HANDLING.
ONE LAYER INVERTING MATTRESS SUTURES INSERTED ALONG FAR SIDE OF ANASTOMOSIS, SIMILAR LAYER BEING INSERTED ALONG NEAR SIDE.
COMPLETED ANASTOMOSIS WILL GIVE ABOUT 90° JOIN – "END TO BACK"

FIG. 5.—Illustrating technique used in 'end-to-back' anastomosis (after Denis Browne, 1951).

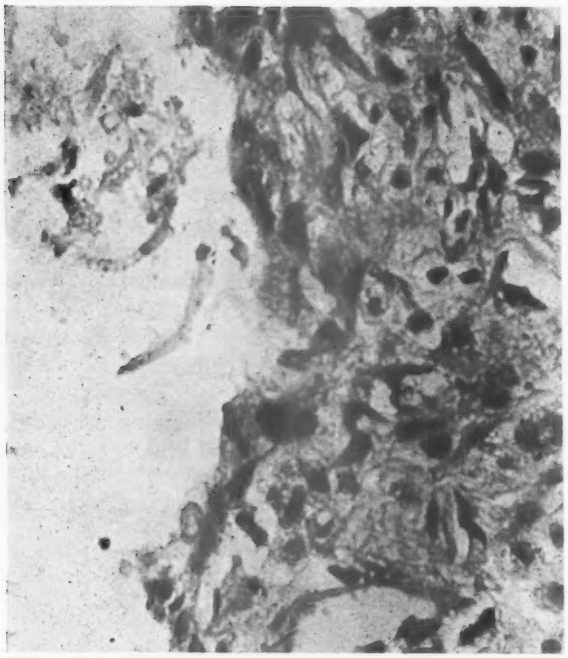

FIG. 6.—Epithelial squames in the lumen of the bowel below a complete atresia. (× 570 approx.)

of Tandler (1900) and Forssner (1907) of persistence of the solid stage of development of the bowel mucosa. But persistence of a mucosal septum is the least common type of atresia in this region. Indeed our histological studies have shown it to be the first

FIG. 7.—Bismuth oxychloride injection of vessels in the region of ileal atresia.

layer to be lost and it is replaced by a loose fibroblastic tissue. Furthermore, other observers such as Johnson (1910) were unable to confirm the presence of such a solid stage in the intestine below the duodenum. Also we have found epithelial squames in the lumen of bowel below a complete atresia (Fig. 6). These have to be swallowed to reach the lumen and the embryo has no such squames on its skin to be swallowed until the third month (Keith, 1948) well after the time of the suggested solid stage. Farber's test is a crude one, and Emery (1952) has recently pointed out that with other staining methods a few squames could be recognized in the meconium of cases of atresia.

Another suggested cause was some vascular accident *in utero*, and Davis and Poynter (1922) described endarteritic obliteration of the arcades

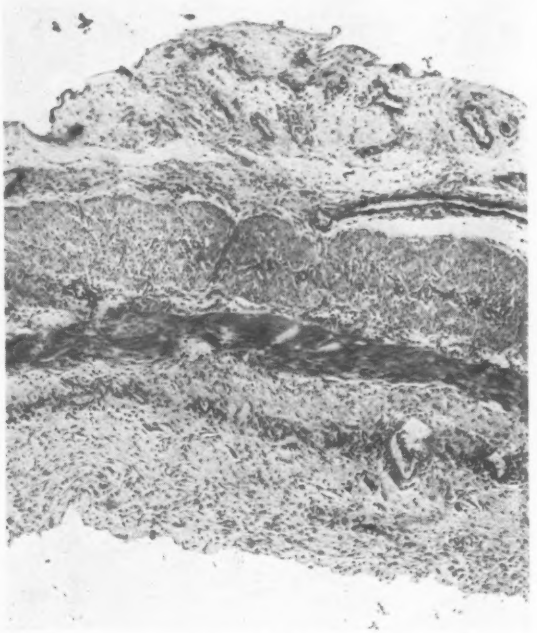

FIG. 8.—A microscopic nodule of meconium peritonitis on the serosa in a case of ileal atresia (also shows loss of mucous membrane). (× 55 approx.)

corresponding to the atretic areas in one case. The vessels of four consecutive cases were injected using a technique based on that of Barlow, Bentley and Walder (1951). I found no abnormality of the feeding vessels beyond a variation in calibre comparable to that of the bowel supplied (Fig. 7).

I suggest that a more likely cause of atresia would be an accident to the bowel after its formation, such as incarceration of the physiological umbilical hernia with resulting nipping or adhesion of parts of the intestine and a predisposition to volvulus of the mass thus formed.

To consider this hypothesis: first, can an accident to formed bowel result in atresia? One typical ileal atresia with a complete gap was found to contain the remnants of an intussusception with a gangrenous apex in its distal bowel. Further examination showed microscopic foci of meconium peritonitis similar to that shown in Fig. 8. The cause of the atresia would appear to be clear in this type of case of which others have been described. Second, does incarceration of the physiological umbilical hernia ever occur? The baby depicted in Fig. 9 was born with an exomphalos into which two open ends of bowel protruded as a spontaneous ileostomy. Around them in the sac lay meconium debris and necrotic remnants of the intervening bowel. Third, is there any evidence of abnormality in the process of reduction of the hernia and replacement of the bowel within the abdomen in these cases? The mesentery was abnormal in nine of the 21 in which it was adequately examined. Also volvulus of a part of the small intestine was present in 11 of the 21 cases. Severe foetal peritonitis was present in five cases and localized adhesions in five more. Microscopic evidence of meconium peritonitis was found in several other cases with no obvious peritonitis. Furthermore, three cases had an associated cystic fibrosis of the pancreas and yet another had long segment Hirschsprung's disease with no ganglia up to the level of the ileal atresia. Both these conditions would produce foetal obstruction and a tendency to dilatation and volvulus of the bowel.

Fig. 10 illustrates a case of volvulus with changes in the wall at the site of twisting, all the layers being lost except the serosa but without actual stenosis.

Fig. 11 illustrates a case of volvulus with severe stenosis of the segment looped around the base. This phenomenon has been observed at either end of the volvulus so that the volvulus cannot be explained as a result of loading of the bowel above a performed atresia.

Fig. 12 depicts an atresia with a cord running between the lumina above and below the lesion. Such cases have shown microscopical meconium peritonitis compatible with nipping in the neck of the umbilical hernial sac causing damage to the bowel wall.

Fig. 13 depicts a complete atresia with a gap between the bowel ends. This type has also shown microscopical meconium peritonitis and some cases have had an isolated segment or segments of bowel between the two ends. Here nipping off at the neck of the sac may have caused complete separation of the bowel and in some cases the intervening segment has grown large enough to persist as a recognizable isolated mass of bowel.

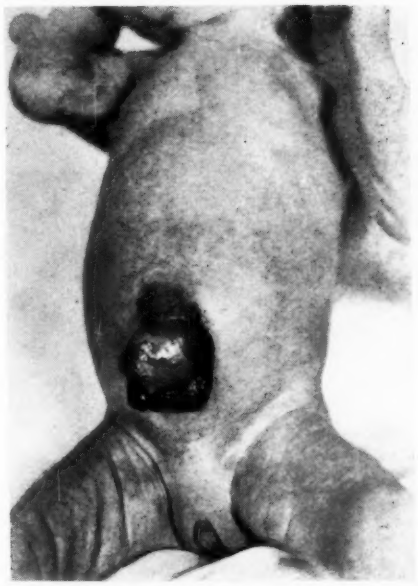

FIG. 9.—Child born with spontaneous double-barrelled ileostomy at umbilicus, with bowel remnants and meconium peritonitis in associated omphalocoele sac.

A mucosal septum is rare in jejuno-ileum, but I have seen two cases outside this series. Again, such a lesion could result from external pressure causing fusion of the mucosal walls.

This concept of a causation of the jejuno-ileal atresias is only a hypothesis, but I suggest that it fits more of the observed facts better than the other hypotheses. As a practical point, such an embryologically late accident to the alimentary tract would be less likely to be associated with serious lesions in other systems. This is borne out in fact; for example, the association with mongolism found in duodenal atresias is not present. The only mongol in these 33 cases was a baby who also had duodenal atresia, and other serious lesions in other systems were absent.

Meconium Ileus

Several cases have now been added to the 14 recently published from The Hospital for Sick Children, Great Ormond Street (Nixon, 1953). All of our cases have had fibrocystic disease of the pancreas, and it seems that inspissation of the meconium is due to the abnormality of the mucus in this disease rather than to pancreatic achylia *per se*.

Fortunately the severity of the intestinal lesion is not necessarily associated with similarly severe pulmonary lesions. The successful case reported is now 4 years old and she is a fit girl who has had no severe respiratory infections. Modern care of this disease can make the saving of a case of meconium ileus well worth while.

FIG. 10.

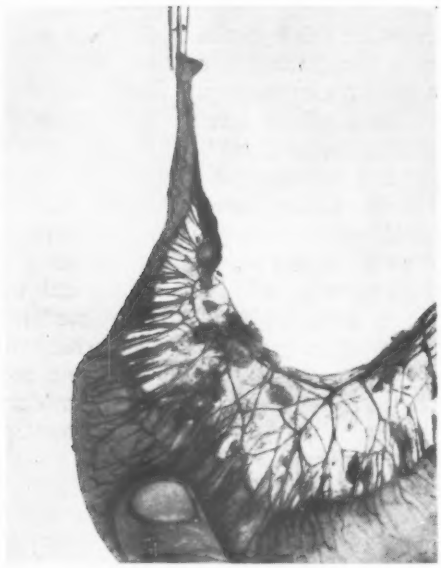

FIG. 12.

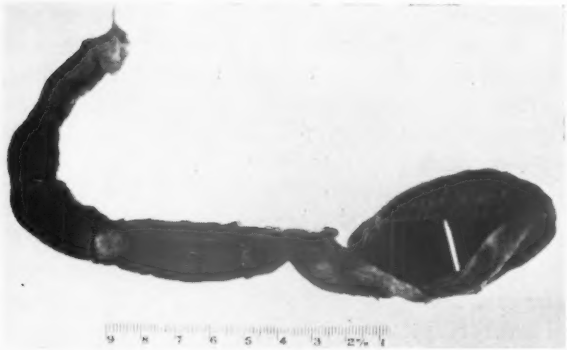

FIG. 13.

FIG. 10.—Lesion in the ileal wall at site of volvulus. Only the serous coat persists.

FIG. 11.—Congenital volvulus of part of small bowel with severe stenosis at site of volvulus shown in original position and after unwinding.

FIG. 12.—Ileal atresia with cord between lumina above and below.

FIG. 13.—Typical hypertrophied and dilated terminal bulb above an ileal atresia. This operative specimen shows the extent of the resection carried out.

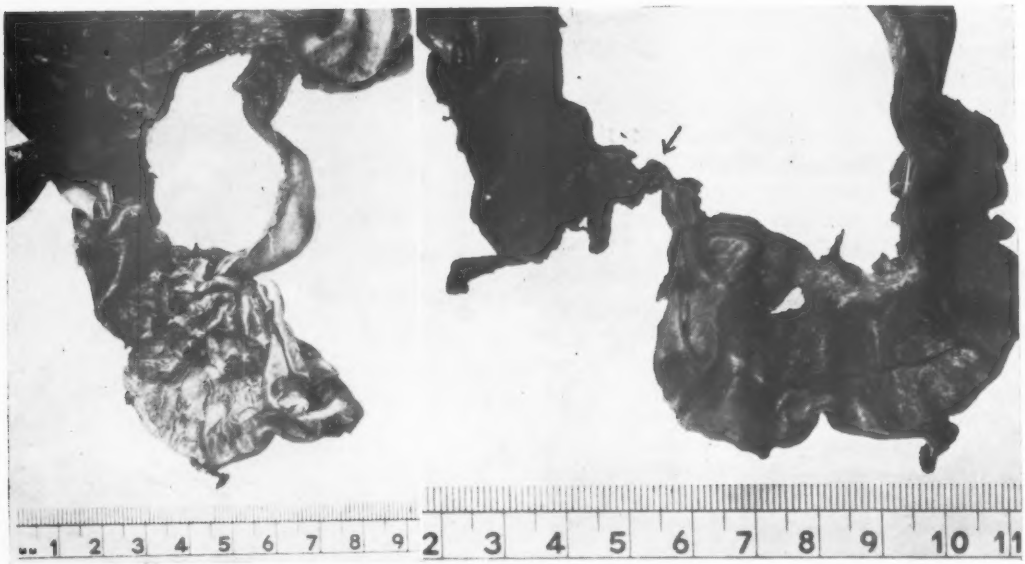

FIG. 11

In my previous report I suggested that the immediate threat to life was perforation of the maximally dilated loop of bowel due to over-distension with, in some cases, an added volvulus. The simplest means of overcoming this risk is an ileostomy which must be above the plug of meconium. Natural infection of the bowel contents will soon liquefy them and pancreatin instillations may assist.

This method was successful in the case mentioned. However, in meconium ileus we are dealing with another form of acute chronic obstruction and the hypertrophied loop may be inefficient in propulsion though viable. Hence resection with a double-barrelled ileostomy as practised by Gross (1953) would seem more reliable.

Many of our cases have had pre-natal volvulus, often with perforation and meconium peritonitis and sometimes an associated atresia. Some of these have seemed to present insoluble problems.

Hirschsprung's Disease

Rapid deflation of the abdomen on rectal examination or washout may make the diagnosis obvious. If not, and the lesion is discovered at laparotomy, then we have found that in this age group the site of the cone of narrowing bowel may be misleading. During foetal life the meconium can be driven down into a long aganglionic segment so that a colostomy may be sited too low in the bowel. The management of this particular problem in Hirschsprung's disease does not seem to be settled. Possibly an ileostomy followed by early re-operation with facilities for examinations of frozen sections may be the answer.

Management in General

Again, I wish only to stress certain particular personal opinions.

I believe that the use of gastric aspiration before and during transfer to a surgical unit would save some of the unfortunate pulmonary inhalation complications which seem so unnecessarily to jeopardize the chances of these babies.

Attempts at pre-operative resuscitation should not be unduly prolonged. Urgent operation is indicated in almost every case. Resuscitation should be started but can be continued during the operation. This sounds unorthodox but the babies do stand up to it. Several times I have seen some accident such as perforation or volvulus occur during the few hours devoted to pre-operative treatment so that the patient has died even before operation could be attempted. If intestinal intubation were practicable in the newborn the urgency would be lessened but gastric intubation is an inefficient means of decompressing the small bowel.

A full laparotomy is almost always used and I prefer a transverse supra-umbilical muscle-cutting incision. It makes prompt atraumatic elucidation of the pathology much easier and avoids the risk of missing the not uncommon second lesion.

Therapy with fluids should be conservative. Blood and plasma for shock and nourishment are life saving. Plasma should be given regularly until oral feeds are adequate to prevent protein depletion. This is a much easier task than to attempt to correct an established depletion. Water should be given sparingly, not more than 40 ml. per lb. per day for maintenance and in an electrolyte solution such as half strength Darrow's solution which will not supply too much sodium chloride. There is a practical danger of giving too much to keep the drip running. The resulting diuresis may get rid of some of the excess water but may also deplete other components of the body fluids not so easily replaced, even if frank drowning by sodium retention does not occur.

Nor is there any need to hurry the oral feeding. The operation soon relieves the obstruction and converts the situation to one of simple starvation which a newborn baby stands well. The aim should be, as Gross puts it, 'to keep the baby on the dry side'.

I would like to thank all the members of the medical and nursing staff of The Hospital for Sick Children, Great Ormond Street, for their cooperation and advice and for allowing me to operate on many of their patients. My particular thanks are due to Mr. Denis Browne and to Dr. Bodian, and to the Research Committee for grants which have enabled me to continue this work. My thanks are also due to Mr. Derek Martin and Mr. P. G. Cull, of the Department of Medical Illustration, for their excellent work.

I also wish to thank the staff of the Alder Hey Children's Hospital, Liverpool, in particular Miss I. Forshall and Dr. E. Hall, for similar help during a period spent in that hospital.

I am indebted to Dr. A. White Franklin for arranging facilities at Queen Charlotte's Maternity Hospital, and to Professor A. Moncrieff for similar facilities at the Postgraduate Medical School.

Professor Slome and the staff of the Department of Physiology of the Royal College of Surgeons of England have given invaluable advice and facilities for the animal work.

Finally, I should like to express my great debt to the late Professor Sir James Spence for many valuable discussions. Although I left Newcastle several years ago, he was never too busy to help.

References

Barlow, T. E., Bentley, F. H. and Walder, D. N. (1951). *Surg. Gynec. Obstet.*, **93**, 657.

Bodian, M., White, L. L. R., Carter, C. O. and Louw, J. H. (1952). *Brit. med. J.*, **1**, 77.

Browne, D. (1951). *Proc. roy. Soc. Med.*, **44**, 623.

Dann, W. J. (1936). *Biochem. J.*, **30**, 1644.

Davis, D. L. and Poynter, C. W. M. (1922). *Surg. Gynec. Obstet.*, **34**, 35.
Dott, N. M. (1923). *Brit. J. Surg.*, **11**, 251.
Emery, J. L. (1952). *Archives of Disease in Childhood*, **27**, 67.
Ehrenpreis, T. and Sandblom, P. (1949). *Acta paediat., Uppsala*, **38**, 109.
Forssner, H. (1907). *Arb. Anat. Inst., Wiesbaden*, **34**, 1.
Frazer, J. E. and Robbins, R. H. (1915). *J. Anat., Lond.*, **50**, 75.
Gross, R. E. (1953). *The Surgery of Infancy and Childhood*, p. 150. Philadelphia and London.
Hoppu, K. (1951). *Ann. Chir. Gynaec. Fenn.*, **40**, Suppl. 1.
Johnson, F. P. (1910). *Amer. J. Anat.*, **10**, 521.
Jolleys, A. (1952). *Brit. J. Surg.*, **40**, 201.
Keith, A. (1948). *Human Embryology and Morphology*, 6th ed., p. 651. London.
Ladd, W. E. (1932). *New Engl. J. Med.*, **206**, 277.
—— (1933). *J. Amer. med. Ass.*, **101**, 1453.
Louw, J. H. (1952). *S. Afr. J. clin. Sci.*, **3**, 109.
Nixon, H. H. (1953). *Brit. J. Surg.*, **41**, 184.
Rickham, P. P. (1952). *Lancet*, **1**, 332.
Santulli, T. V. (1954). *J. Pediat.*, **44**, 317.
Spencer, R. (1951). *Brit. J. Surg.*, **39**, 105.
Swain, V. A. J. and France, N. E. (1954). *Lancet*, **1**, 844.
Tandler, J. (1900). *Morph. Jb.*, **29**, 187.

PERITONITIS IN THE NEONATAL PERIOD*

BY

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It has generally been assumed that peritonitis occurs but rarely in newborn infants. In the 1952 Charles West lecture (Moncrieff, 1953) on infection in newborn babies the condition was not mentioned. Thelander, who wrote a classical paper on perforations of the gastro-intestinal tract in newborn infants in 1939, stated that most paediatricians never saw a case, and in 1949 Low, Cooper and Cosby could only find 100 instances in the literature since 1761, when Morgagni first reported the condition in *De Sedibus et Causis Morborum*. During the last five years, however, many cases of peritonitis in infants have been described and it has become clear that a condition which was formerly thought to be a pathological curiosity, is in fact a not uncommon cause of disease and death in the neonatal period. The pathology and aetiology are, as yet, little understood and recent publications have caused confusion by the synonymous use of terms, such as meconium peritonitis, intra-uterine peritonitis, foetal peritonitis, and peritonitis in the newborn.

This paper is presented in order to describe our experience with neonates suffering from peritonitis, to evaluate the incidence of the disease, and to classify the various types of the condition. It is based upon the case histories of 17 newborn infants seen by us during the last three and a half years. During this period we admitted about 250 babies to the Neonatal Surgical Unit at Alder Hey Children's Hospital; 98 of these had intestinal obstruction (excluding pyloric stenosis and imperforate anus). It appears, therefore, that about 7% of all newborn infants admitted for urgent surgical conditions and 17% of all cases of neonatal intestinal obstruction are suffering from peritonitis.

Of the 17 cases under discussion, seven were classified as meconium peritonitis, and 10 as acute bacterial peritonitis. There was a marked preponderance of girls, 12, or 70%. This is surprising as it is generally stated (Table 1) that perforation of the intestinal tract during the newborn period affects boys three times more commonly than girls (Thelander, 1939).

Physiological and Pathological Considerations

Meconium Peritonitis. It is generally accepted that meconium starts to accumulate in the intestine of the 4-month-old embryo, but there is some evidence that the 3-month-old foetus is already swallowing amniotic fluid and passing it along the

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

TABLE 1

RESULTS IN PRESENT SERIES

Name	Age	Weight	Finding at Operation	Operative Procedure	Result
Meconium Peritonitis:					
Zena	10 hrs.	6 lb. 9 oz.	Volvulus of jejunum	Resection and anastomosis	Recovery
John	3 days	5 lb. 2 oz.	Ileal atresia	,, ,,	Died 14 days later
Hilary	2 days	6 lb.	Meconium ileus	,, ,,	Died 12 hr. later
Linda	3 days	5 lb. 2 oz.	Perforation of ileum	,, ,,	Died three days later
Brian	3 weeks	9 lb. 12 oz.	Ileal obstruction due to bands	Division of bands	Recovery
Sarah	3 weeks	6 lb.	Ileal obstruction due to bands	,, ,,	Recovery
Joan	2 days	5 lb. 6 oz.	Jejunal atresia	Resection and anastomosis	Died four days later
Acute Bacterial Peritonitis:					
Mary	2 weeks	6 lb. 9 oz.	Abscess round terminal ileum	Drainage and ileocolostomy	Recovery
Nancy	2 weeks	5 lb. 14 oz.	Abscess in R. iliac fossa	Drainage of abscess	Recovery
Stephen	8 days	3 lb.	Perforation of stomach	Closure of perforation	Died four days later
Jean	1 day	5 lb. 7 oz.	Gangrenous volvulus of ileum	Resection and anastomosis	Recovery
Lesley	5 days	5 lb. 3 oz.	Perforation of ileum	,, ,,	Recovery
Loraine	3 weeks	6 lb. 8 oz.	Pelvic abscess	Drainage of abscess	Recovery
Doreen	3 days	2 lb.	Perforation of Meckel's diverticulum	Resection and anastomosis	Died three days later
Alexander	3 weeks	7 lb. 9 oz.	Abscess in R. iliac fossa	Drainage of abscess	Recovery
Angela	1 week	6 lb.	Perforated duplication of ileum	Resection and anastomosis	Died four weeks later
Michael	4 weeks	7 lb. 9 oz.	Perforated ileum	Suture of perforation	Recovery

gastro-intestinal tract (Davies and Potter, 1946). According to Keith (1933), it reaches the ileo-colic junction at four months and the rectum at five months of intra-uterine life. Meconium peritonitis can therefore theoretically occur during the last six months of pregnancy and Rudnew (1915) reported the condition in a 6-month-old foetus. Once meconium escapes into the peritoneal cavity it produces a chemical peritonitis. Numerous adhesions form, which, on microscopy contain meconium nodules and meconium-laden giant cells.

Two years ago we reported three cases of meconium peritonitis (Forshall, Hall and Rickham, 1952). We then agreed with Boikan (1930) in restricting the term 'meconium peritonitis' to those cases where 'meconium, calcified meconium, mucous droplets, foreign body giant cells, cells from the site of perforation, fibrous and fibrinous adhesions, and rarely lanugo hair, can be demonstrated in the peritoneal cavity'. Bendel and Michel (1953) defined meconium peritonitis rather similarly as a 'non-bacterial foreign body and chemical peritonitis occurring during intra-uterine or early neonatal life as a result of an abnormal communication between the bowel contents and the peritoneal cavity.' If the abnormal communication is still patent after birth the original sterile chemical peritonitis will soon become infected and a secondary pyogenic peritonitis will result. We do not think that these cases should be included under the title 'meconium peritonitis' as the clinical picture and the operative and necropsy findings are those of acute bacterial peritonitis, the presence of meconium being incidental.

Meconium peritonitis can be divided into two groups on an aetiological basis (Ramos, 1950). Group 1 includes all cases with intestinal obstruction. First, the cause of the obstruction can be in the lumen of the gut, i.e., meconium ileus, as in our Case 2, and in the cases described by Kornblith and Atani (1929) and Porter and Weeks (1915). Secondly, it can be in the wall of the gut, either an intestinal atresia, as our Cases 2 and 7, and numerous others reported in the literature (Neuhauser, 1944; Low *et al.*, 1949; Chandler, 1949) or, less commonly, an intestinal stenosis (Butler *et al.*, 1945). Thirdly, the cause of the obstruction may be outside the gut, for example, a volvulus as in our Case 1, and in cases described by Abt (1931) and Sturzenegger (1927), or it can be due to extrinsic congenital bands (Farr and Brunkow, 1925).

Group 2 comprises all those cases where there is perforation of the bowel wall, without distal obstruction. We have seen three such cases (Cases 4, 5 and 6) and numerous other examples can be found in the literature (Franklin and Hosford, 1952). There has been speculation as to the cause of intestinal perforation in the absence of obstruction. Davis and Poynter (1922) pointed out that perforation is by no means common, even in cases with intestinal atresia; the back pressure due to the intestinal block does, however, offer some explanation for a rupture of the thinned wall of the proximal intestine. In cases without intestinal obstruction one has to assume that there is an abnormality in the wall of the gut. Aplasia of the muscularis mucosa has been suggested as the cause of these perforations by von Sury (1912) and Moretti (1949), while Helbing (1908) and Paltauf (1888) postulated a primary vascular insufficiency of the affected intestine. Finally, Boikan (1930), Lattes (1943), and Maguire and Moore (1950) have brought forward evidence that a marked hypertrophy of the glands of Lieberkuhn, coupled with thinness of the intestinal wall and lymphoid hyperplasia, may be a predisposing cause of perforation. This would explain why the lower ileum is the common site of perforation as in our Case 4.

In many of the reported cases of meconium ileus no sign of a perforation can be found at operation and one has to assume that the perforation seals itself off during intra-uterine life (Peiser, 1908; Davis and Poynter, 1922; Flesch, 1925). In six of our seven cases no perforation could be demonstrated. We were, however, able to show by serial section that there were deep ulcers with complete destruction of the muscular layers in the proximal distended segment of the gut, and that in Case 1, which had a volvulus, the walls of the involved gut showed incomplete layers of musculature which were replaced by granulation tissue containing giant cells and calcified meconium. It is therefore reasonable to assume that in many instances there is a perforation *in utero* which closes before birth.

The question of trauma as an aetiological factor in the causation of meconium peritonitis has been discussed very fully in the literature. Rupture of the gut due to trauma usually occurs during and after birth, and although some meconium may exude into the peritoneal cavity, bacterial peritonitis will quickly supervene. We have seen only one case where trauma was the cause of peritonitis in the newborn, and nowadays such cases of mismanagement are rare. Russell (1928) reviewed the cases reported to be caused by trauma. Most of them were described in the nineteenth century by German workers (Zillner, 1884; Paltauf, 1888) and at that time the medico-legal aspect of the condition was widely discussed. Lee and MacMillan (1950)

in a critical review of the published cases doubted if trauma were an aetiological factor in many of them.

Whatever the cause of the entrance of meconium into the peritoneal cavity, the effect is always the same, namely, a plastic peritonitis with numerous adhesions binding down the intestine. These adhesions may cause kinking of the intestine and hence obstruction, as shown in two of our patients (Cases 5 and 6).

Acute Bacterial Peritonitis. Under this heading we have included all those cases where a gastro-intestinal perforation occurred during or after birth and where no meconium was found in the peritoneal cavity. It is well known that acute peritonitis can occur in early infancy without gastro-intestinal perforation. This is usually due to blood-stream infections with such organisms as streptococcus, gonococcus or pneumococcus (Gubern Salisachs, 1951). Peritonitis due to *Bact. coli* is very rarely seen as a complication of gastro-enteritis. Such cases have become uncommon and only rarely come to the surgeon's notice.

Four of our nine cases came first to our notice when a large intra-peritoneal abscess had formed in the right iliac fossa or the pelvis (Cases 8, 9, 13 and 15). At operation, no attempt was made to demonstrate the site of perforation, and as all the infants survived and subsequent barium follow-through studies did not reveal any abnormality, we are not certain if any part of the gut ruptured. It might have been a perforated lower ileum, a Meckel's diverticulum, or an appendix, or due to blood-stream infection, although the age, clinical picture and localization of the abscesses are rather against this hypothesis. In the other six cases, the site of rupture was seen at operation. We found a rupture of the lesser curvature of the stomach, a perforation of a gangrenous volvulus, two ruptures of the lower ileum, a perforated Meckel's diverticulum and a perforation in an ileal duplication respectively.

In discussing acute bacterial peritonitis in the newborn, the following aetiological factors have to be considered:

PERFORATED PEPTIC ULCER. Perforation of peptic ulcers have not infrequently been encountered during the newborn period. Bird, Limper and Mayer (1941) collected from the literature 42 cases of peptic ulcers in newborn babies. Duodenal ulcers were twice as frequent as gastric ones. The presenting symptom was either bleeding or perforation. In 1953 Greene and Gose collected 20 reported cases of perforated ulcers and added two more which had been under their care. The great Cruveilhier is often credited with having described the first case in 1829, but Siebold (1825) reported a perforated peptic ulcer in a newborn infant three years previously. The first successful operation for this condition was performed by Léger, Ricard, Léonard and Piette in 1950. Since then Kellogg, Abelson and Cornwell (1951), Brink and Keyser (1952), and Moncrief (1954) have reported further successes. Perforation of a gastric ulcer *in utero* was described by Lee and Wells (1923). Several theories about the aetiology of peptic perforations have been advanced: Wright and Scott (1950) thought that ulcers might develop because of the extraordinary high acidity of the stomach contents during the first few days of life. The gastric acidity reaches adult values during this period (Miller, 1941). Herbut (1943) postulated a defect in the gastric musculature and Russell (1940) considered trauma to be an aetiological factor. In Case 10 of our series there was a large rent in the lesser curvature which had caused gross pneumoperitoneum (Fig. 1). The obstetric history of this 3 lb. premature baby is of interest. There was a prolapse of the cord and the child was delivered as a difficult breech delivery at home. No cause for the perforation was found at operation and the defect was closed by sutures. The baby died four days after operation and at necropsy an extensive tear in the falx and massive extradural haemorrhage

FIG. 1.—Case 10: radiograph of abdomen, child erect, showing gross pneumoperitoneum.

were found. The immediate cause of death was a thrombosis of the inferior vena cava causing infarction of the liver, spleen, stomach and duodenum. It is likely that the original tear in the lesser curvature was due to the same vascular catastrophe. Our experience with this baby is of interest in view of Guthrie's (1942) theory that the anoxia associated with prolonged labour leads to devitalization of the duodenal mucosa. Our case certainly had a difficult and prolonged delivery. Intracranial haemorrhage in association with perforation of a gastric ulcer has also previously been reported by Gottlieb, Chu and Sharlin (1950).

ACUTE APPENDICITIS. Acute appendicitis is a great rarity in this age group. In 1945 Etherington-Wilson could only discover five cases in the literature and reported a sixth. He overlooked a case reported by Ch'eng and K'ang (1937). All these children died shortly after birth. Since then one more fatal case (Creery, 1953) and one case surviving operation (Meigher and Lucas, 1952) have been reported.

PERFORATED MECKEL'S DIVERTICULUM. Perforation of Meckel's diverticulum in the neonatal period has occasionally been described. Ungari and Valiani (1952) had a fatal case where the perforation was due to ectopic gastric mucosa. Another fatal case was described by Rosza and Gross (1953).

FIG. 2.—Case 17: rupture of small omphalocoele. The protruded small intestine is strangulated and black.

In Case 14 of our series there was a small gangrenous area near the base of the diverticulum which had perforated. Normal intestinal mucosa was found on section.

PERFORATED INTESTINE. Perforation of the small or large intestine is the cause of the majority of cases with peritonitis in the newborn. As mentioned before, the lower ileum is the site most frequently involved. In our Cases 11 and 17 such a perforation was found and in Cases 8, 9, 13 and 15 it is possible that the peritonitis was due to a similar cause. In considering the aetiology of these ruptures, Gross and Ferguson (1952) have pointed out that the gastro-intestinal tract in newborn infants shows a greater development of its secretory and absorbing surfaces than of its musculature. In Case 11 there was a marked thinning of the intestinal wall around the site of perforation. Defects in the muscular coat of the intestine have been considered as the cause of some neonatal intestinal perforations, especially when associated with increased intestinal pressure due to partial obstruction (Qvigstad, 1950).

Trauma as a cause of perforation of the intestine has already been discussed under meconium peritonitis. In Case 17 of our series trauma may have been an aetiological factor.

This baby was perfectly well until he was 8 days of age, when the shrivelled remnants of the umbilical cord dropped off. It was then noticed that a loop of bowel protruded through a small defect at the umbilicus. By the time the child was admitted to our unit, the whole of the small intestine had prolapsed. It was black and appeared not viable (Fig. 2). As it was obviously impossible to resect the whole length of intestine, it was returned to the abdomen and the defect was repaired. To our surprise, the child made an uninterrupted recovery and was discharged 10 days after operation. Two and a half weeks later the child was readmitted as an emergency with a history of vomiting for 12 hours. On examination he was very shocked and cyanosed, the respiration was grunting and the abdomen was enormously distended and silent. Some mucus was passed per rectum. A radiograph of the abdomen revealed free gas under the diaphragm. At operation the abdominal cavity was found to be filled with faeces and pus and numerous dense adhesions had formed between the coils of intestine. There was a rent, half an inch long, in the wall of the lower ileum. The rent was sutured transversely and the abdomen closed. A biopsy from the edge of the intestinal defect revealed no evidence of specific infection or thrombosis on microscopy. The child made a good recovery.

In retrospect, it seems possible that after the first

FIG. 3.—Case 16: perforation in a duplication of the ileum. The probe points to the site of perforation.

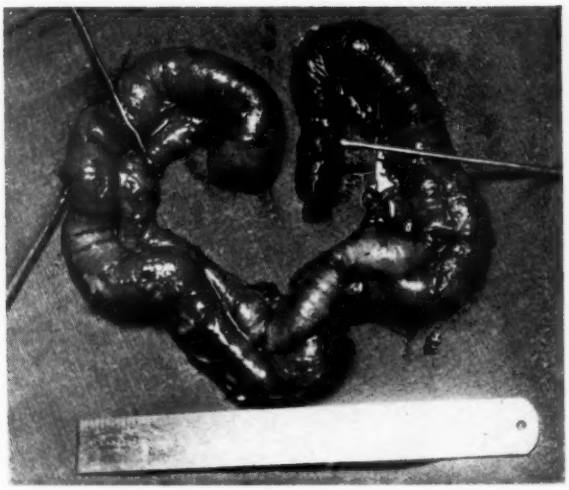

FIG. 5.—Case 16: the excised segment of ileum with duplication. The perforation can be seen on the right side.

operation the partially devitalized area of gut became surrounded by adhesions. Before the second operation these adhesions may have been ruptured by minimal trauma, such as lifting the child or compressing the abdomen.

Perforation of a duplication of the intestinal tract is another rare cause of acute bacterial peritonitis. Only one of the 68 cases of duplication reported from the Boston Children's Hospital had perforated. Resection was performed and the child died (Gross, 1953). We have quite recently seen a case with a duplication of the lower ileum about 1 ft. in length. The proximal end of the duplication ended blindly, while the distal end opened into the main lumen of the ileum. There was a round perforation of about 3 mm. in diameter (Figs. 3 to 6). The child, a girl weighing 5 lb. 9 oz., had been quite well for one week after birth. She then started vomiting and

FIG. 4.—Case 16: the mesentery of the ileum and its duplication have been divided. The perforation is marked by the probe.

FIG. 6.—Case 16: the anastomosis between the proximal and distal part of the ileum has been completed.

six hours later collapsed; she was resuscitated. Eighteen hours after the onset of the illness straight radiographs of the abdomen showed multiple fluid levels. There was no gas under the diaphragm. She was admitted to our unit 24 hours after the first vomiting. On admission she was very shocked and toxic with a high temperature. The abdomen was distended and there was definite tenderness in the right iliac fossa. A provisional diagnosis of bacterial peritonitis, probably due to intestinal perforation, was made. At operation, the whole length of the ileum with its duplication was resected and an end-to-end anastomosis was performed.

PYOGENIC PERITONITIS. Finally, pyogenic peritonitis may occur secondary to gangrene of the bowel as in our Case 10, in which there was a gangrenous volvulus of the mid-ileum; a similar case was described by Arnheim (1945).

BILE PERITONITIS. This is only mentioned in order to complete the picture. It is excessively rare; I could only find one case reported in the literature (Byrne and Bottomley, 1953). The peritonitis was due to a ruptured choledocus cyst.

The aetiology of peritonitis in the newborn is summarized in Table 2.

Diagnosis

The signs and symptoms of meconium peritonitis are those of intestinal obstruction in the newborn. In cases associated with atresia of the gut, the clinical picture will develop shortly after birth, but in cases where the obstruction is due to strictures or bands symptoms might not come on for some time. Two patients in our series, Cases 5 and 6, had no trouble until they were 3 weeks old. Bile-stained vomit, abdominal distension and constipation are the presenting symptoms and their severity depends upon the site and completeness of the obstruction.

With a patent intestinal perforation at birth, the symptoms of acute bacterial peritonitis will soon supervene. Shock, cyanosis, rapid, grunting respiration, abdominal distension, bile-stained vomit, oedema of the flanks and scrotum, and occasionally in the absence of obstruction, the passage of blood and mucus per rectum, are described by Thelander (1939). Not all these symptoms may be encountered in the same patient, but some are always present. Shock is very marked when the perforation is recent, especially in small babies. Cases 10 and 14 in our series, weighing 3 lb. and 2 lb. respectively, were good examples of infants suffering from profound shock. Abdominal distension with or without visible peristalsis, was observed to a greater or lesser extent in all our babies. Rapid, grunting respiration and cyanotic attacks are usually secondary to abdominal distension causing pressure on the diaphragm, but may also be due to inhalation of vomit. In three children, Cases 10, 12 and 17, absence of the liver dullness due to pneumoperitoneum was noticed. Oedema of the flanks and genitalia was observed in four infants, Cases 8, 9, 13 and 15. In two of them, the oedema was associated with a blotchy erythema of the abdominal wall. We have not encountered a neonate suffering from peritonitis who passed blood per rectum, but Cases 13 and 17 passed quantities of mucus before the pelvic abscess was evacuated through the rectum. In the last four cases under discussion, the infection localized as a large intraperitoneal abscess and a mass could be felt in the right iliac fossa in two cases. The palpation of this mass clinched the diagnosis.

Radiology can help in the diagnosis of these conditions and a straight radiograph of the abdomen

TABLE 2

PERITONITIS IN THE NEONATAL PERIOD

- (1) Meconium Peritonitis
  - *Group* 1 with intestinal obstruction
    - (i) In lumen of gut, i.e., meconium ileus
    - (ii) In wall of gut, i.e., atresia, stenosis
    - (iii) Outside gut, i.e., volvulus, hernia, bands
  - *Group* 2 without intestinal obstruction
    - (i) Defect in muscularis
    - (ii) Vascular accident
    - (iii) Hypertrophy o glands of Lieberkuhn
- if perforation patent after birth → (2) Acute Bacterial Peritonitis
  - (i) Acute appendicitis
  - (ii) Perforated Meckel's diverticulum
  - (iii) Perforated peptic ulcer
  - (iv) Perforation of intestine
  - (v) Gangrenous bowel
  - (vi) Trauma
  - (vii) Bloodstream infection
  - (viii) Infected haemoperitoneum
  - (ix) Transmural infection
- (3) Bile Peritonitis

FIG. 7.—Case 12: perforation of the ileum. The radiograph in the erect position shows air under the diaphragm.

often reveals the nature of the condition. Kirchhoff in 1932 was the first to describe a pneumoperitoneum when radiographing a newborn baby. It is obvious that only cases with a patent intestinal perforation will show free gas in the abdomen. Three of our cases were diagnosed on this radiological finding (Figs. 7 and 8).

In meconium peritonitis the extruded meconium frequently calcifies within the peritoneum. Litten (quoted by Boikan, 1930) stated that this calcification can occur within 24 to 48 hours. This phenomenon can be reproduced experimentally by introducing meconium into the peritoneal cavity of rats (Rubovits, Taft and Neuwelt, 1938). Calcified masses can occasionally be observed in radiographs of the abdomen (Fig. 9). This diagnostic radiological sign was first recognized by Neuhauser in 1944. As we have previously pointed out (Forshall, Hall and Rickham, 1952), calcification is often so slight that it cannot be seen on a straight abdominal radiograph, although it can be demonstrated in x-ray films of the excised tissues.

Rarely, intra-abdominal calcification is discovered accidentally in babies not suffering from intestinal obstruction. Olnick and Hatcher (1953) report the case of a baby of 2 months of age whose radiographs showed calcification in the abdomen and scrotum. The diagnosis was confirmed by scrotal biopsy.

Calcification may occasionally occur in meconium within the lumen of the bowel, the peritoneum being unaffected (Camp and Roberts, 1949). The differential diagnosis can only be made at operation.

A straight radiograph of the abdomen may also be helpful in those cases where a mass can be felt on palpating the abdomen. In Case 15 of our series there was some doubt if a palpable mass was an enlarged, displaced kidney. An intramuscular pyelogram showed normally functioning kidneys and a soft tissue shadow displacing the gas-filled intestine (Fig. 10).

Treatment

The treatment of peritonitis in the newborn is the treatment of the underlying condition. This is not the place to discuss treatment in any detail, as it would involve discussing the management of all types of intestinal obstruction in the neonatal

FIG. 8.—Case 14: perforated Meckel's diverticulum. The radiograph of the abdomen in the erect position shows a small air bubble between the liver and the diaphragm.

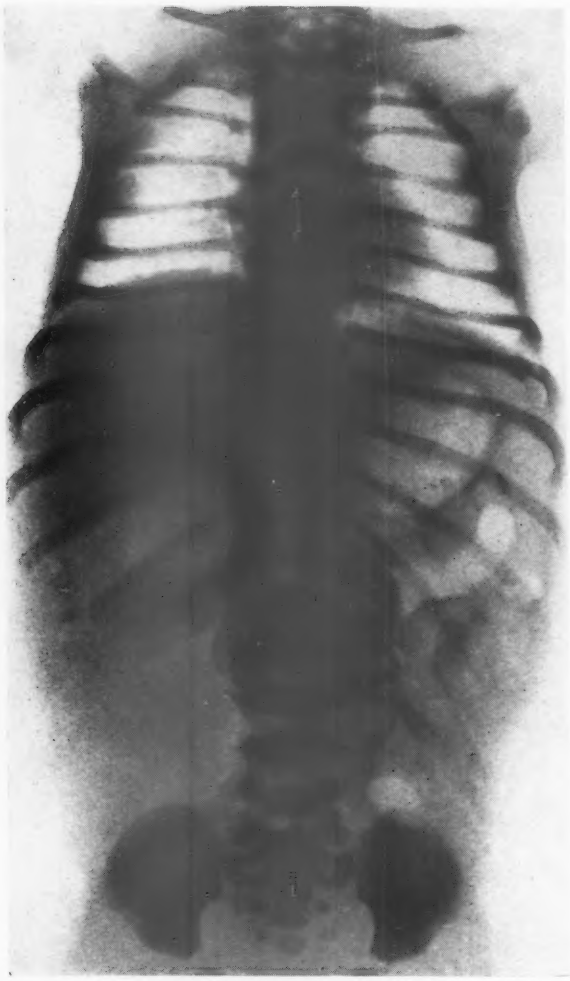

FIG. 9.—Case 7: Straight radiograph of the abdomen showing intra-abdominal islands of calcification.

period. It is hardly necessary to stress that shock, dehydration and electrolyte disturbances must be corrected if these small infants are to survive operation. Most of them need intravenous therapy and continuous gastric suction for some hours pre-operatively until their blood chemistry has come back to normal and their general condition has improved. At operation, bands or adhesions should be divided if they cause demonstrable obstruction. Gastric perforations should be closed by suturing the defect, but when encountering intestinal perforations, we prefer resection of the affected loop of gut and end-to-end anastomosis. Intestinal atresia, or stenosis, should also be treated by wide resection of the affected segment and end-to-end anastomosis. The numerous intraperitoneal adhesions may prevent a thorough inspection of the distal intestine and produce subsequent intestinal obstruction, but we did not attempt to separate these bands at operation as it was felt that this would greatly increase operating time, produce a considerable amount of bleeding and shock, and lead to the formation of new adhesions. We have usually thought it wiser to close the abdomen after resection and anastomosis had been completed. In those cases surviving operation, no further obstruction developed.

Where there is a large intraperitoneal abscess, this should be drained by the most convenient route. The abscess wall should not be disturbed and we believe it is unwise to search for the site of perforation. There is, of course, danger of a faecal fistula developing subsequently if this treatment is employed. One of our four cases of abscess developed such a fistula post-operatively; this closed spontaneously after a week. The three cases which were treated according to this principle had a very satisfactory convalescence. In Case 8 no mass was felt pre-operatively and the baby was operated upon for intestinal obstruction of unknown origin. At operation, an abscess surrounded by coils of lower ileum was found in the right iliac fossa. The

FIG. 10.—Case 15: radiograph of the abdomen showing a constant shadow in the right iliac fossa which displaces the gas-filled intestine. It was found at operation to be an intra-peritoneal abscess in the right iliac fossa.

abscess was drained, but the densely adherent loops of gut were still obstructed. Rather than dissect out the adhesions, it was felt that a short-circuiting side-to-side ileo-transverse colostomy offered the best chances for survival. The child made an uninterrupted recovery. She has been followed up for over three years and has no further symptoms.

Post-operatively, a prolonged period of ileus is to be expected. Intravenous therapy, continuous low-pressure gastric suction and vigilant control of any changes in the blood chemistry will, in our experience, overcome this dreaded complication.

Mortality

In this series of 17 cases there were seven deaths, a mortality of 41·2%. This high figure compares unfavourably with a mortality of 19·8% in 81 cases of intestinal obstruction without peritonitis in newborn babies operated upon during the same period of three and a half years. It appears, therefore, that peritonitis in the newborn carries an extra risk and is, in our experience, next to meconium ileus, the most fatal of the intra-abdominal catastrophes in this age group.

The causes of death were as follows: three children died of bronchopneumonia. One of them had, in addition, a congenital malformation of the heart and another was very premature, weighing only 2 lb. The causes of death in the other four were gastro-enteritis, overwhelming toxaemia, thrombosis of the inferior vena cava and intestinal fistula producing inanition. In retrospect, two of these deaths might have been prevented, if at the time we had known more about the correct post-operative management of these infants.

In conclusion, we should again like to stress that peritonitis in the newborn is not a very rare condition. It is still frequently not diagnosed during life and only discovered at necropsy. In a survey of neonatal deaths in Liverpool during 1949 several records of such cases were found (Rickham, 1952). Once diagnosed, the condition is curable by operation and should be associated with a reasonable chance of survival.

I should like to thank the paediatricians of the Liverpool region who referred these cases to us and whose vigilance in detecting surgical emergencies in newborn babies has played such a big part in lowering our operative mortality.

I should like to acknowledge Miss I. Forshall's help in preparing this paper. We collaborated closely in treating these infants and Miss Forshall performed half the operations.

References

Abt, I. A. (1931). *Med. Clin. N. Amer.*, **15**, 611.
Arnheim, E. E. (1945). *Amer. J. Dis. Child.*, **69**, 108.
Bendel, W. L. and Michel, M. L. (1953). *Surgery*, **34**, 320.
Bird, C. E., Limper, M. A. and Mayer, J. M. (1941). *Ann. Surg.*, **114**, 526.
Boikan, W. S. (1930). *Arch. Path., Chicago*, **9**, 1164.
Brink, E. ten and Keyzer, J. L. (1952). *Maandschr. Kindergeneesk.*, **20**, 108.
Butler, A. M. (1945). *New Engl. J. Med.*, **233**, 257.
Byrne, J. J. and Bottomley, G. T. (1953). *Amer. J. Dis. Child.*, **85**, 694.
Camp, R. and Roberts, M. H. (1949). *Ibid.*, **78**, 393.
Chandler, L. R. (1949). Discussion of paper by C. W. Brunkow *et al.* *West. J. Surg.*, **57**, 424.
Ch'eng, Y. H. and K'ang, H. J. (1937). *Chin. med. J.*, **52**, 876.
Creery, R. D. G. (1953). *Brit. med. J.*, **1**, 871.
Cruveilhier, J. (1829-42). *Anatomie Pathologique du Corps Humain.* Paris.
Davies, M. E. and Potter, E. L. (1946). *J. Amer. med. Ass.*, **131**, 1194.
Davis, D. L. and Poynter, C. W. M. (1922). *Surg. Gynec. Obstet.*, **34**, 35.
Etherington-Wilson, W. (1945). *Proc. roy. Soc. Med.*, **38**, 186.
Farr, R. E. and Brunkow, C. W. (1925). *Arch. Surg., Chicago*, **11**, 417.
Flesch, H. (1925). *Jb. Kinderheilk.*, **108**, 366.
Forshall, I., Hall, E. G. and Rickham, P. P. (1952). *Brit. J. Surg.*, **40**, 31.
Franklin, A. W. and Hosford, J. P. (1952). *Brit. med. J.*, **2**, 257.
Greene, W. W. and Gose, D. F. (1953). *Amer. J. Dis. Child.*, **85**, 47.
Gross, R. E. (1953). *Surgery of Infancy and Childhood.* Philadelphia and London.
—— and Ferguson, C. C. (1952). *Surg. Gynec. Obstet.*, **95**, 631.
Gottlieb, C., Chu, F. and Sharlin, H. S. (1950). *Radiology*, **54**, 595.
Gubern Salisachs, L. (1951). *Acta pediá. esp.*, **9**, 27.
Guthrie, K. J. (1942). *Archives of Disease in Childhood*, **17**, 82.
Helbing, T. (1908). *Ueber foetale Peritonitis, nebst einem casuistischen Beitrag aus der Universitäts. Frauenklinik zu Freiburg.* Inaug. Diss., Freiburg.
Herbut, P. A. (1943). *Arch. Path., Chicago*, **36**, 91.
Keith, A. (1933). *Human Embryology and Morphology.* 5th ed. London.
Kellogg, H. G., Abelson, S. M. and Cornwell, F. A. (1951). *J. Pediat.*, **39**, 357.
Kirchhoff, H. (1932). *Röntgenpraxis*, **4**, 233.
Kornblith, B. A. and Atani, S. (1929). *Amer. J. Path.*, **3**, 249.
Lattes, R. (1943). *Amer. J. Obstet. Gynec.*, **46**, 149.
Lee, C. M. and MacMillan, B. G. (1950). *Surgery*, **28**, 48.
Lee, W. E. and Wells, J. R. (1923). *Ann. Surg.*, **78**, 36.
Léger, J. L., Ricard, P. M., Léonard, C. and Piette, J. (1950). *Un. méd. Can.*, **79**, 1277.
Low, J. R., Cooper, G. and Cosby, L. (1949). *Surgery*, **26**, 223.
Maguire, C. H. and Moore, W. R. (1950). *Surgery*, **28**, 568.
Meigher, S. C. and Lucas, A. W. (1952). *Ann. Surg.*, **136**, 1044.
Miller, R. A. (1941). *Archives of Disease in Childhood*, **16**, 22.
Moncrieff, A. (1953). *Brit. med. J.*, **1**, 1.
Moncrief, W. H. (1954). *Ann. Surg.*, **139**, 99.
Moretti, I. (1949). *Minerva pediat., Torino*, **1**, 239.
Morgagni, J. B. (1761). *De Sedibus et Causis Morborum.* Typographia Remondiniana, Venice.
Neuhauser, E. B. D. (1944). *Amer. J. Roentgenol.*, **51**, 421.
Olnick, H. M. and Hatcher, M. B. (1953). *J. Amer. med. Ass.*, **152**, 582.
Paltauf, A. (1888). *Virchows Arch. path. Anat.*, **111**, 461.
Porter, L. and Weeks, A. (1915). *Amer. J. Dis. Child.*, **9**, 283.
Peiser, A. (1908). Bruns' *Beitr. klin. Chir.*, **60**, 168.
Qvigstad, I. (1950). *Nord. Med.*, **43**, 504.
Ramos, J. F. (1950). *J. nat. med. Ass.*, **42**, 105.
Rickham, P. P. (1952). *Lancet*, **1**, 332.
Rosza, S. and Gross, R. J. (1953). *Amer. J. Roentgenol.*, **69**, 944.
Rubovits, W. H., Taft, E. and Neuwelt, F. (1938). *Amer. J. Obstet. Gynec.*, **36**, 501.
Rudnew, W. (1915). *Ueber die spontanen Darmrupturen bei Foeten und Neugeborenen.* Inaug. Diss., Basel.
Russell, T. H. (1928). *J. Amer. med. Ass.*, **90**, 1931.
—— (1940). *Trans. New Engl. Surg. Soc.* (1939), **22**, 286.
Siebold, A. E. von (1825). *J. Geburtsh.*, **5**, 3.
Sturzenegger, E. (1927). *Beitr. path. Anat.*, **78**, 85.
Sury, K. von (1912). *Vjschr. gerichtl. Med.*, **43**, II Supplement-Heft, 91.
Thelander, H. E. (1939). *Amer. J. Dis. Child.*, **58**, 371.
Ungari, C. and Valiani, A. (1952). *Clin. Ostet. Ginec.*, **54**, 147.
Wright, L. T. and Scott, B. E. (1950). *J. Pediat.*, **37**, 905.
Zillner, E. (1884). *Virchows. Arch. path. Anat.*, **96**, 307.

ACUTE INTUSSUSCEPTION IN CHILDHOOD*

BY

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Although there are many aspects of intussusception which would well repay close consideration, there are two serious problems that frequently confront the children's surgeon—the management of a gangrenous intussusception and the role of conservative treatment.

In my experience death during or shortly after operative treatment of intussusception has usually been due to inadequate prevention or treatment of shock. Resuscitative measures have become such a routine and often successful procedure in the severely ill child that the mere institution of these measures may come to be regarded as adequate preparation for operation. A repeated assessment of the condition of a seriously ill child must be made by the surgeon, and it is important to resist the temptation to operate early for one's own convenience.

How long should resuscitative treatment be continued? If the infant improves rapidly there is no problem, but some will need three to six hours before they should be submitted to laparotomy. Still others, desperately ill, may not in fact show any clear sign of recovery and, in spite of this, they must be operated upon. I do not think a definite time limit should be placed on resuscitative treatment, but any worthwhile improvement will usually occur within the first six hours.

A second difficulty is to decide how long to spend on the attempt at reduction. Some surgeons would put a limit of 15 or 20 minutes. Although it is true that one may lose all sense of time during the uneasy persuasion of a recalcitrant intussusception, I think it is a mistake to limit the attempt by the clock but rather by the progress of reduction. There are times when it is apparent after three or four minutes that reduction is impossible. On other occasions a difficult reduction may take a long time but the bowel be found healthy on completion.

If it has been decided that an intussusception is irreducible, or if the bowel is found to be gangrenous after reduction, there is no doubt about the need for resection, but there is disagreement about operative technique.

It is sometimes said that the simplest and safest procedure is to suture the afferent and efferent loops of bowel together to make a spur and bring the mass out of the abdomen, where its removal leaves a double-barrelled enterostomy. While it is true that this can be done quickly, the post-operative course is by no means always smooth. In the first place the loss of a considerable volume of intestinal fluids may be an extremely grave matter even with careful fluid therapy and although the spur is crushed after three or four days. Moreover, the closure of the double-barrelled enterostomy is not without its own risks.

The second method is enterostomy with anastomosis, in which an anastomosis is made between the afferent and efferent loops at the time of the first operation. Another variation in this technique is to perform an anastomosis, perhaps an end-to-side or side-to-side anastomosis and lead a catheter from the proximal end through a stab wound as a safety measure.

The third type of procedure is a straight resection and anastomosis without any enterostomy as a safeguard. I think there are two important aspects in this operation. The first is that the resection should be generous and extend well beyond the oedematous bowel. The second point is the advantage of deflating the proximal bowel if it is grossly distended. My own preference is for the closed end-to-end anastomosis where this is possible, but I think it is sufficiently clear that the open anastomosis fails, when it does fail, not because of some incidental operative contamination of the abdominal cavity but because the technique of intestinal suture is at fault or the condition of the intestinal wall is poor.

My choice of technique for the gangrenous intussusception would be influenced by my assessment of the child's condition before operation and the advice of the anaesthetist about his progress or deterioration during the procedure. Were I not satisfied that the patient was reasonably fit I would

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

bring out the affected loop and remove it when the abdomen was closed. In other cases resection and end-to-end anastomosis is preferred.

The second important problem in the management of intussusception is the vexed question of conservative treatment. Ever since Jonathan Hutchinson in 1871 first reduced by operation an intussusception in a child there has been a strong tradition of operative treatment in this country and in America. At the same time as Hutchinson (1874) reported his success, Hirschsprung in Copenhagen was systematically treating all cases by an enema coupled with manipulation through the abdominal wall, and by 1905 he was able to present 107 cases treated in this way. In Australia, Clubbe (1921) was most insistent that 'the treatment of intussusception is laparotomy', but when the patient was on the operating table under anaesthesia he always tried an olive oil enema. In 140 cases, 10% were reduced and in many more the apex was pushed back as far as the caecum. The lead that Clubbe had given in Australia was followed there by Hipsley, who in 1926 published a report of 100 cases in which a saline enema was used on the operating table. He claimed to assess reduction by measuring the girth of the abdomen after evacuating the enema. If it had increased there must be fluid in the small bowel and the intussusception was reduced. He reported 62 out of 100 reduced in this way.

The non-operative procedure received a great stimulus in 1927, when George Retan in America described the use of the barium enema in reducing an intussusception. His suggestion was taken up with enthusiasm in Scandinavia, where Nordentoft (1943) in Denmark reported 440 cases in which 31% were reduced. In Sweden Hellmer published his experiences in 1948, in which he claimed that in a series of 162 cases, 80% of the intussusceptions were reduced.

Criticisms of Conservative Treatment

I had given little consideration to the conservative approach to intussusception, nor was I tempted to try it after reading the paper of Ravitch and McCune (1948a) in which the results of operative and conservative treatment were compared. The fallacy of this comparison had been recognized by Hutchinson, who wrote:

> 'It is absurd to institute any comparison between the treatment by injection and that by laparotomy. Statistical tables indicating a contrast as regards the relative success of the two measures are a mere waste of labour.'

However, I was encouraged to give the methods a trial as a result of a very reasoned reply which I had received from T. Y. Nelson of Sydney, after I had criticized his paper in *The Medical Journal of Australia* (1949). The criticisms of the conservative treatment which have been levelled from time to time fall under six headings: (1) Delay, (2) perforation, (3) uncertainty of reduction, (4) missing a high intussusception, (5) missing a causative lesion, and (6) a high rate of recurrence.

Delay. It is said that delay before operation entailed by the attempt to reduce the intussusception with barium enema is serious and, moreover, that the procedure itself is shocking. However, provided the facilities and staff are available, the attempt can be completed within an hour of admission to hospital, by which time the pre-operative atropine has become effective. Also one of the difficulties about the conservative method is that when the intussusception has been reduced as far as the caecum the child may seem somewhat better and the surgeon must not be deceived by the improvement in general condition into thinking that reduction is complete. If the attempt were made, and I do not advocate it, in the seriously ill child it might well be that the child's condition would be worse, not as a result of the delay, but because of the failure to apply resuscitative measures.

Danger of Perforation. Several cases of perforation have been recorded, but all that I have heard of have been late cases. In one of their papers Ravitch and McCune (1948b) described some experiments in which they showed that in dogs a pressure of three feet of water was unable to cause perforation in a gangrenous intussusception and provided the pressure is kept no higher than this, the chances of perforation appear minimal. If perforation does occur, in the words of Nyborg (1943) 'the prognosis is pessima'.

Uncertainty of Reduction. The certain sign of reduction is to observe the barium pass high up into the small bowel under the fluorescent screen, but it cannot always be persuaded to pass beyond the ileocaecal valve. In such cases, the complete filling of the caecum and the absence of a mass on palpation after evacuation have been accepted as reasonable security. If doubt exists about the reduction the abdomen must be opened.

High, Purely Ileal Intussusception. The criticism that a high, purely ileal intussusception may be missed is based on a misconception of the purpose of the barium enema in intussusception. It is not intended as a diagnostic measure. If the clinical diagnosis is intestinal obstruction due to intussus-

ception and a barium enema fails to show the intussusception the abdomen must be opened.

Missing a Causative Lesion. Lesions such as a polyp, Meckel's diverticulum, tumour or enterogenous cyst may be the leading point of an intussusception and at operation they can usually be recognized and dealt with at the time if the conditions are favourable, or removed later. It is true that a lesion of this sort might be missed with reduction by means of a barium enema and on this score the criticism cannot be adequately answered.

High Rate of Recurrence. The usual figures given for recurrence in intussusception following operation are somewhere between 2 and 5%. After conservative reduction it is said to be high, and Hellmer, in his series of 162 cases, reported a remarkably high incidence of 16%. In my own small series I have not been able to confirm this high incidence. If there is recurrence it is likely to be recognized easily and early and the case can again be submitted to a barium enema.

Technique

The two opposing views in the past have been, first that operation should be undertaken at once if the child is fit, and secondly that every case should have an attempted reduction with a barium enema. It seemed to me possible that the truth might lie somewhere between these two extremes and that a further study of the problem was justified.

In view of the criticisms a determined attempt was made to avoid as many pitfalls as possible and yet, at the same time, maintain simplicity. The house officers were given some rules to avoid the dangers of delay and shock and the child was always given pre-medication with atropine so that he would be ready for the theatre if necessary, and rectal thiopentone sodium was given as a sedative. The barium was warm and a douche can was held in the hand so that the pressure could not be too great. The buttocks were held by a volunteer and the barium run in slowly.

The intussusception may be met in the descending colon and pushed back as far as the transverse colon but no further (Fig. 1). On the other hand it may be completely reduced and the barium flow high into the small bowel (Fig. 2). Finally, it may pass as far as the caecum and a filling defect be obvious (Fig. 3).

FIG. 1(*a*)—The barium enema has outlined the intussusception in the descending colon.

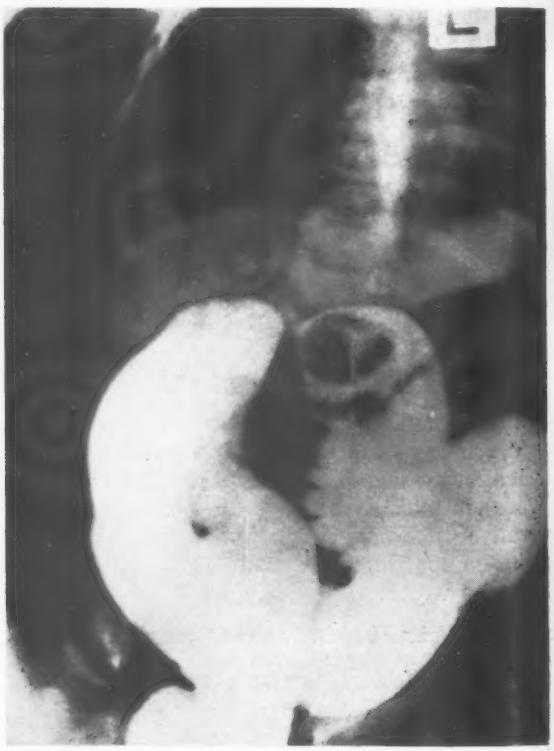


FIG. 1(*b*)—Reduction took place as far as the transverse colon but no further.

FIG. 2(*a*)—The intussusception was outlined by the barium enema in the transverse colon.

FIG. 2(*b*)—Barium has passed high up into the small bowel and the intussusception is reduced.

FIG. 3.—Reduction as far as the caecum but filling defect still present.

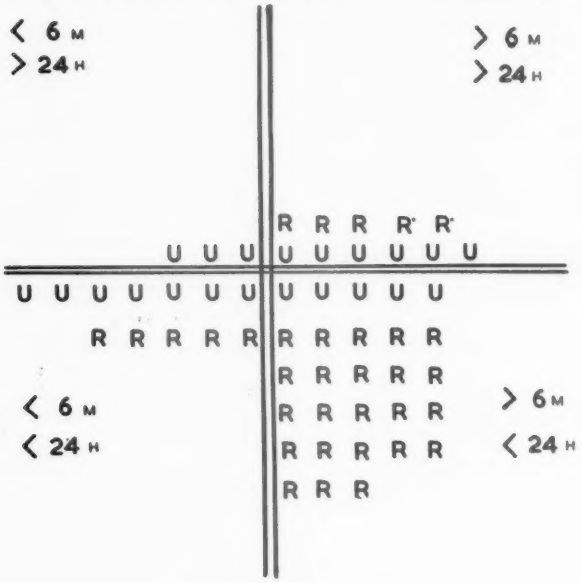
Results

In 54 cases there was an attempt at reduction with barium enema and in 33 it was successful. Among the factors which might be influencing the success or failure of the procedure delay before admission to hospital seemed the most likely. When the delay was less than 24 hours, 28 out of 40 were reduced (70%), whereas when the delay was more than 24 hours, only three out of 12 (25%) were reduced, excluding two cases in which the intussusception had been present for more than a week in older patients who were not acutely ill.

It also seemed possible that the intussusception would be tighter in a young child, simply because the bowel was smaller. Among children of less than 6 months of age only five out of 15 (33%) were reduced, whereas in those more than 6 months of age 28 out of 39 (72%) were reduced. If these two factors are combined it is found that among those children who are more than 6 months of age with a delay of less than 24 hours, 23 out of 28 intussusceptions were reduced, approximately 82%. These results can be represented graphically (Fig. 4).

FIG. 4.—Chart to show influence of time and age factors.

R = Intussusceptions reduced by barium enema.
U = Intussusceptions not reduced by barium enema.
R· = Chronic case—delay more than one week.
< 6 M., > 6 M. = Age less than or more than 6 months.
<24 H., >24 H. = Delay less than or more than 24 hours.



Conclusions

With selection of cases, successful reduction of intussusception with barium enema can be achieved in a high proportion.

If the technique is restricted to those admitted within the first 24 hours there is little danger.

Perhaps most important of all, the procedure should be in responsible hands and the child must be under the direct care of the surgeon who may have to operate.

Summary

The infant with a gangrenous intussusception needs adequate resuscitative treatment before and during operation. For the experienced surgeon the treatment of choice will be resection and anastomosis if the child is fit, or a double-barrelled enterostomy if his condition is desperate. Conservative treatment has a place in the management of the early case provided that adequate facilities and experienced staff are available.

This study of the conservative reduction of intussusceptions was made possible by the very kind collaboration of Dr. T. Lodge, Consultant Radiologist of the Children's Hospital, Sheffield, not only by offering the facilities of his department but also by his personal supervision of the radiological reduction in most of the cases.

REFERENCES

Clubbe, C. P. B. (1921). *The Diagnosis and Treatment of Intussusception*, 2nd ed. London.
Hellmer, H. (1948). *Acta radiol., Stockh.*, Suppl. 65.
Hipsley, P. L. (1926). *Med. J. Aust.*, **2**, 201.
Hirschsprung, H. (1905). *Mitt. Grenzgeb. Med. Chir.*, **14**, 555.
Hutchinson, J. (1874). *Med. chir. Trans.*, **57**, 31.
Nelson, T. Y. (1949). *Med. J. Aust.*, **1**, 825.
Nordentoft, J. M. (1943). *Acta radiol., Stockh.*, Suppl. 51.
Nyborg, S. (1943). *Acta chir. scand.*, Suppl. 80.
Ravitch, M. M. and McCune, R. M. (1948a). *Ann. Surg.*, **128**, 904.
—— —— (1948b). *Bull. Johns Hopk. Hosp.*, **82**, 550.
Retan, G. M. (1927). *Amer. J. Dis. Child.*, **33**, 765.

CONGENITAL DEFORMITIES OF MECHANICAL ORIGIN*

BY

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This and the following paper are both on subjects belonging to that strangely neglected group of human ills, the congenital deformities. To compare this group with others of similar importance, for instance infections or new growths, is to find that there is no generally accepted classification of its elements, and no textbook treating of them. What information is available has to be extracted from papers scattered through the literature and a few monographs; there is no general review of the subject.

The first lecture indeed treats of a group of deformities which is not generally admitted to exist at all, those due to mechanical faults in the mechanically extremely complicated process of gestation. It is strange that though it is universally admitted that mechanical faults can have their obvious consequences in the cavities of the skull or the chest, it is almost universally denied that similar processes and results may occur in the cavity of the uterus. One explanation of this may be that the experimental method, on which modern medicine relies almost entirely, is of little use here: it is too difficult to alter intra-uterine conditions without bringing the pregnancy to an end. In consequence the chief method available is one much used in such scientific pursuits as astronomy, that of working out in the imagination what would happen were certain hypotheses true, and then comparing these abstract conclusions with what actually occurs. The difference from the experimental method, in which actual changes are made in the course of events (for instance, Duraswami's researches on the effect of insulin on the developing egg), is fundamental, so that it is all the more strange that there is no term in common use to describe and distinguish this method of using the mind.

Abnormal Mechanics in Pregnancy

It is possible to imagine the process of gestation going wrong mechanically in three main ways:

Malposition. By this term I mean that the developing foetus, which must obviously be tightly folded up, is folded up in the wrong way. Here one comes against another interesting gap in present teaching; the lack of any generally accepted view on how the foetus is normally folded. Keith (1948), for instance, teaches that the feet lie *in utero* with their soles against each other, but this does not explain the usual excess of movement into calcaneus that a newborn baby's foot possesses over that of an adult. If we presume that the normal intra-uterine position is with the soles of the feet against the uterine wall and, of course, with the hips, knees, and elbows flexed, then this hypothesis is confirmed by the fact that the range of movement in the joints of a newborn infant corresponds to what would be expected from a prolonged sojourn in this position. That is, the elbows, knees, and hips will not extend to the adult range, and the feet go up into calcaneus much further, so that the little toes can easily be made to touch the outer side of the leg.

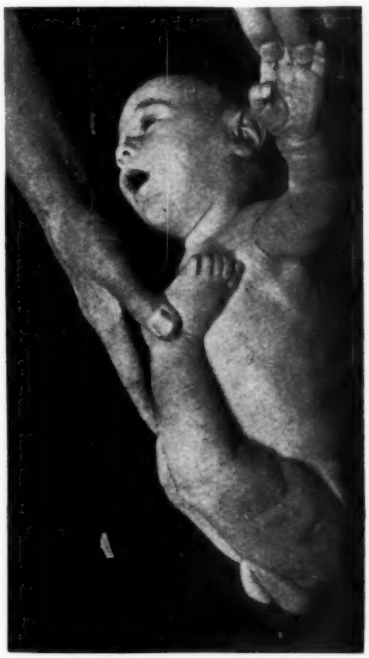

FIG. 1.—Example of malposition *in utero*. The pregnancy was normal in size and sensations, and the baby showed no evidence of excessive pressure. The only abnormality is that the knees are extended instead of flexed.

I do not know any orthopaedic work which comments on these facts, which is perhaps to be accounted for by most orthopaedic authorities at present being committed to a dogmatic denial

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

that pathological conditions of the feet and joints can have mechanical causes. To admit that in the newborn there are normal variations from the adult state of obvious mechanical origin would be to allow the entry of the thin end of a heretical wedge-shaped idea.

Increased Mechanical Pressure. It might be assumed that increased pressure on a limb would have the same effects on the tissues before birth as it would have afterwards. If a plaster case is put too tightly on a limb the effects are two: atrophy of the muscles and stiffening of the joints. Probably the main cause of these effects is interference with the venous return, but the explanation of the stiffening of the joints is not obvious.

It might also be expected that if there were undue tightness of the uterus on the foetus, which should be the only cause of this pressure, there would be two effects felt on the maternal side. (1) The mother's abdomen would be smaller than in normal pregnancies. (2) There would be discomfort for the mother, particularly noticeable when she has had a normal pregnancy to compare with the abnormally tight one.

On checking this hypothesis with what actually occurs, it is found to be constantly confirmed: that is to say that children showing evidences of pressure in atrophied muscles and stiff joints emerge from small and uncomfortable pregnancies. The legs and feet are, of course, much more affected than the arms, which are sheltered by the large overhanging foetal head.

Increased Hydraulic Pressure. An increase in the tension of the amniotic fluid might be expected to interfere with the venous returns from the limbs just as a similar increase in the intracranial tension spoils the venous return from the retina and causes optic atrophy. The difference in the effects of this pressure from those of the mechanical type would be that owing to the laws of hydraulics it would be equally distributed over all four limbs. Its effects would also be expected to be more severe as the distance from the pump of the foetal heart becomes greater, that is to say more in the feet than in the hips, and more in the hands than in the shoulders.

FIG. 2.—Example of excessive mechanical pressure. The pregnancy was a very small and uncomfortable one. The feet and legs have been in the normal position, but have been moulded to conform closely to the concavity of the uterus. The joints of the legs are stiff, and the muscles have hardly developed at all.

FIG. 3.—Example of increased hydraulic pressure. The pregnancy was a very large and uncomfortable one, with excessive amniotic fluid. The fixed stiff extension of the hips could only occur in a uterine cavity larger than normal. Note also the cylindrical limbs, showing none of the normal moulding of the tissues over the bony points, owing to lack of contact with the uterine wall. The joints are stiff, and the muscles undeveloped, equally in all four limbs owing to the laws of hydraulics, a contrast to the normal arms in Fig. 2.

Increased hydraulic pressure suggests that usually there would be an abnormally large amount of fluid: though, of course, it would be possible to have increased hydraulic pressure with a normal or abnormally small amount. Compare again the situation in the cranial cavity, where increased cerebral tension may exist either with the increased amount of fluid of hydrocephalus, or the decreased amount found in cerebral tumours.

Here again the maternal histories of cases of 'arthrogryposis' confirm the hypothesis. The mothers give a story of abnormal pain of the kind associated with abnormal hydraulic pressure in the skull, the bladder, or a joint. A very large proportion of these children emerge from hydramniotic pregnancies, and they often show the joints of the limbs stiffened in positions which would only be possible in an abnormally large cavity: for example, the hips are extended instead of flexed in the normal way.

It is characteristic of the attitude of present-day

medicine towards this subject of congenital deformities that the correlation of abnormal babies with abnormal pregnancies is almost entirely neglected. The reason is that the obstetrician is very little interested in the baby, and the paediatrician equally uninterested in the details of pregnancy, while the orthopaedic surgeon is hampered by the dogmatic attitude which I have already mentioned.

Classification of Deformities due to Mechanical Conditions

Deformities of the Feet. There are three regular deformities which occur continually in different degrees of the same moulding.

METATARSAL VARUS. This condition is due to mal-position with the soles of the feet together. There is also a form which is combined with metatarsal valgus of the other foot in a mutual deformity, which is assumed to the feet being diverted sideways in the normal position.

CLUB-FOOT, EQUINO-VARUS OR TOTAL VARUS. I object to the name equino-varus as in the more severe degrees the feet are in calcaneus, and I suggest the name 'total varus' to describe this condition in which the ankle and heel are moulded inwardly as well as the forefoot. The deformity can be most shortly described by saying that it is exactly what would result were one to press the outer side of the model of a foot in putty against a concavity corresponding to that of the uterine wall.

METATARSAL VALGUS. This is not due to mal-position, but to increased mechanical pressure exaggerating the normal calcaneus position of the newborn foot so that it wraps itself round the outer side of the leg. Its association with congenital dislocation of the hip and weak muscles is mentioned later.

In addition to these regular types of deformity there is an infinite variety of irregular forms of moulding, corresponding to various abnormal positions of the limbs. These are associated in particular with spina bifida in those cases in which very early pressure has both interfered with the fusing of the edges of the spinal groove and forced the cerebellum and medulla down through the foramen magnum into the deformity known as the Arnold-Chiari malformation.

It is of interest to compare with the mechanical hypothesis the explanations usually given of feet mouldings of the type vaguely known as talipes.

That they are due to arrest at some stage of development: no one could possibly accept this hypothesis who is familiar with the infinite variety of mouldings of the feet, though at one stage there is a vague likeness between the inversion of the foetal feet and the mildest form of 'equino-varus' talipes (in the severer forms it has been noted that the feet described in this way are in calcaneus, not equinus).

That the deformities are due to the action of abnormal or abnormally attached muscles: apart from the fact that many of the mouldings could not possibly be produced in this way there is the awkward fact that some of the most severe of them occur in children whose muscles can never have been active, cases of spina bifida and arthrogryposis.

That club-foot or total varus is due to a congenital dislocation of the astragalo-scaphoid joint (Brockman, 1930): it is of course impossible to mould a foot into this shape without dislocating slightly this joint as well as others. But this explanation leaves unaccounted for the moulding of the heel and the atrophy of the calf muscles, quite apart from its offering no explanation of the immense variety of other malformations of the same type.

That 'the deformity is not a result of the position, but the position is a result of the deformity': This is a purely verbal objection, as can be seen by transposing it into the statement that 'the deformity of bow-legs is not a result of the position of bow-legs, but the position of bow-legs is a result of the deformity of bow-legs'.

The interesting experiments of Duraiswami (1952) have been accepted in some quarters as suggesting a pathology of talipes and congenital dislocation of the hip. Apart from the fact that such conditions often occur in perfectly healthy children, the deformities produced in chickens have only the vaguest resemblance to those found in man. The resemblance is much more a matter of words than of structure.

Deformities of the Joints. The only joint to be found dislocated in otherwise normal children is the hip, and it is the only joint that could be dislocated by mechanical means given the intra-uterine conditions. Also it is always dislocated in the only direction in which it could be forced, that is to say backwards and downwards by pressure on the knee; and the head and neck of the femur are compressed and anteverted in the way in which a soft structure like the foetal head of the femur would be moulded by being forced out of the socket while the joint is in the intra-uterine position.

The only other joint to be found dislocated, the knee, is only so malformed in association with the malposition of hyperextension, the connexion being very obvious.

Deformities of Other Parts. With the exception of the Arnold-Chiari deformity which has already been briefly mentioned, these are infinite in variety and irregular in type, very often being associated with embryological faults of formation such as missing digits and bones.

An important part of the argument is the combination of different deformities. To take a few of them:

In double talipes the deformity is never exactly equal, but always greater on one foot than the other. This would be expected as the outer foot in the cross-legged position responsible would get more moulding pressure than the inner one.

Congenital dislocation of the hip is constantly associated with valgus talipes. The foot deformity is assumed to be due to increased mechanical pressure, which would also tend by its action on the knee to push the head of the femur out backwards.

Congenital dislocation of the hip is never, in my experience, associated with a straightforward varus moulding of the foot. This is because the pressure on the outside of the foot swings the knee into abduction, which brings the head of the femur forward and keeps it in the acetabulum.

Metatarsal valgus talipes, assumed to be due to increased mechanical pressure, is always associated with weak muscles, which would also be expected from the same cause.

The existence of 'mutual deformities' in which the convexity of an irregular talipes of one foot fits into the concavity of an irregular talipes of the other. It seems to me that the occurrence of such cases would be alone sufficient to prove the moulding hypothesis.

Treatment

The essential idea of the classical treatment which I was originally taught was to treat deformities rather as if they had been cement or plaster, that is to say they were corrected as far as possible, tendons being freely cut to allow this, and they were then immobilized with the idea that they should set in the position obtained. This notion ignores the fact that the body is a vital moving structure, which only develops properly if its parts are being used in the way for which they are fitted. I found its results to be so disappointing that I sought for new principles, and found them in the idea that certain movements which tend to correct the deformity should be permitted, and others which increase it should be prevented. To do this it was necessary to work out a new system of splinting, the main idea of which was to control the system of levers, which is formed by bones, joints and muscles, by means of a counter-balancing system of levers in the form of splints.

The disadvantage of the old method was that the joints stiffened and the muscles atrophied under immobilization and the effects of cutting tendons round the ankle were much the same as those of cutting them round the wrist.

The advantages to be expected from the system of controlled movement would be:

(1) Rapidity of correction, due to the continual manipulations supplied by the automatic actions of the infant; (2) stimulation of the growth of imperfectly developed parts, a stimulation that use alone could give; (3) the acquiring of correct muscle balance by use in the corrected position. Immobilization, of course, simply weakens all the muscles equally, without changing their original faulty relationships of tone.

I have worked upon the idea that to reverse the moulding of the feet one should imitate the kind of pressure that has originally produced it. This pressure has certain characteristics:

(1) It is constant, so that manipulations at intervals do not reproduce it. (2) It is fluctuating, varying *in utero* with the motions of the child and of the uterus. This fluctuation can be imitated by a mechanism that allows movements, even though the movements are selected. One very important result of these fluctuations in pressure is that they tend to pump blood through the tissues, and so prevent that stasis of the circulation which is a main factor in pressure sores, caused either by immobilization in plaster or by prolonged lying still in bed. (3) It is soft and distributed over a wide area. This can be imitated by such material as sticking plaster or adhesive felt. (4) It is frictionless. In the uterus the amniotic fluid prevents friction. In splinting this can also be done by stopping the movement of the material which is applying pressure, that movement which is the essence of friction and consequent soreness, by making this material adhesive. It is quite an important point in technique which is often not understood, that if one uses adhesive felt to apply pressure it should be stuck to the foot, and not to the splint.

It is, of course, impossible to describe here the various forms of apparatus devised for the various deformities, but in my opinion the hopes I held of this method of treatment have been very fairly realized. Of course, some varieties of these deformities are utterly intractable; nothing can produce muscles which have failed to form, and I have never had any real success in diminishing the stiffness of congenitally stiff joints.

It is worth recording, not as a complaint but as

an observation of an instance of a universal human reaction, that I had the utmost difficulty originally in getting my heretical ideas of pathology and treatment published. This was because the editors of journals submitted my articles to the opinion of orthopaedic authorities, who, finding their teachings assailed, naturally said that the attack should not be allowed to go forth to corrupt the young.

At present my main trouble is due to another universal reaction, that of modifying a method or a mechanism in order to establish the superiority of the modifier over the original inventor. It is still apparently difficult for many surgeons to understand the system of levers of which my splints consist, as is shown by the curiously common cutting off of the extremely important outside lever in my 'hobble splint' for talipes.

BIBLIOGRAPHY

Bell, J. F. and Grice, D. S. (1944). *J. Bone Jt Surg.*, **26**, 799.
Brockman, E. P. (1930). *Congenital Club-foot*. London.
Burns, B. H. and Ellis, V. H. (1937). *Recent Advances in Orthopaedic Surgery*, p. 280. London.
Duraiswami, P. K. (1952). *J. Bone Jt Surg.*, **34B**, 646.
Jergesen, F. H. (1943). *Ibid.*, **25**, 185.
Keith, A. (1948). *Human Embryology and Morphology*, 6th. ed. p. 622. London.
Kuhlmann, R. F. (1954). *Amer. J. Dis. Child.*, **87**, 440.
Stamm, T. T. (1948). In *British Surgical Practice*, vol. 4, p. 146, ed. Carling, E. R. and Ross, J. P. London.
Thomson, S. A. (1942). *J. Bone Jt Surg.*, **24**, 291.
Thorek, M. (1949). *Modern Surgical Technique*, 2nd ed., vol. 2, p. 1343. Philadelphia.

CONGENITAL DEFORMITIES OF THE ANUS AND THE RECTUM*

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One of the regular methods of progression in medicine is by the analysis of large vaguely assorted groups of cases into smaller and exactly defined categories. The process consists in a mixture of observation, abstract reasoning and experiment.

As instances of this the work of Hamilton Russell (1922) may be quoted, when by a combination of observation and abstract reasoning he established the existence of inguinal hernias due to a congenital malformation, and distinguished them from those due to purely mechanical causes, which had been classed with them. Then there is the work of Swenson and Bill (1948), which has split up the vague group classed under the unhappy name of megacolon into two classes, one being that of Hirschsprung's disease, a congenital deformity consisting in the absence of ganglion cells in the bowel, and the other a functional failure to empty a normal bowel which can be called 'colonic inertia'. In the same way the splitting up of the group of 'undescended testicles' into 80% of normal variations in development, the retractile class, and 20% of congenital deformities, has made it possible to give definite prognoses and indications for treatment where both were lacking before. It is the purpose of this lecture to suggest that a similar process may with advantage be applied to the heterogenous group known by the name of imperforate anus.

We have found that two conceptions have helped greatly in classification and, far more important, in treatment. These conceptions are those of the ectopic anus, a normal sphincter mechanism which has become displaced as a testicle in its descent may take the wrong line; and the covered anus, in which a normal mechanism has been buried by a process not usually recognized, that of excessive fusion of embryological structures. This failure of development, whose existence was suggested to me by Douglas Stephens, is, of course, the reverse of the well recognized one in which fusion is deficient, with the result of a hare-lip or similar deformity.

Other groups of deformities, those of the various imperfect and stenotic anuses, though recognized, are only recognized by few, and are hardly described at all in textbooks.

The Imperfect Anus

Stenosis of the Anus. The normal anus of the newborn should take the male adult little finger without difficulty, and it may be mentioned that the process of testing this capacity is probably the best treatment for mild degrees of constipation in the small baby. The more severe degrees of stenosis are obvious enough if looked for, though they may escape this investigation for months, with the grave danger of producing the obstinate condition of colonic inertia through loss of the normal irritability of a rectum constantly overloaded with faeces. A less obvious degree consists in a fibrous ring inside the junction of skin and mucosa, which is not

FIG. 1.—Stenosis of anus (microscopic anus). This is the type frequently described as an imperforate anus with merely a diaphragm over the opening. Actually careful examination shows a tiny opening, which merely needs dilatation.

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

apparent on the outside, though it is very obvious to the examining finger.

Microscopic Anus. Though this condition is merely an extreme degree of stenosis, its existence is so little realized and it is so easy to overlook that it merits a separate description. In it the opening from skin to bowel is so minute that its only sign is the occasional emergence of a small spot of meconium, resembling an ordinary 'fly speck'. The important point about these microscopic openings is that they indicate the existence of a proper sphincter mechanism surrounding them. I have known several cases in which quite unnecessary colostomies were performed in ignorance of this.

Treatment. Both the foregoing conditions simply need dilatation, though in the case of the microscopic variety there may be difficulty in finding a probe fine enough to start the process. Successive calibres of sounds should be passed till a normal expansion has been gained. It is best to have the baby in hospital for this unavoidably uncomfortable process, which should be begun under an anaesthetic, and continued daily till the sound passes without difficulty or pain. The child should then be sent out accompanied by the sound and the mother instructed in the use of it. It must be strongly emphasized that in many of these anal conditions long-continued dilatation is indispensable for successful treatment. The passage of the lubricated sound should be continued for several months, and inspection for several months after that.

The Ectopic Anus

The Shot-gun Perineum. In this condition a normal anus and vagina lie with their edges touching, without the normal strip of skin intervening. Usually there is a dimple behind the anal opening to mark the normal site for this. The condition needs no treatment, as appearance is of no great importance and function is unimpaired. In fact most cases go through life unaware of their departure from the normal, and an eminent senior obstetrician tells me that he remembers demonstrating such a case as 'congenital absence of the perineum' in a woman who had successfully produced several children.

FIG. 2.—Ectopic anus (shot-gun perineum). The anus has moved forward so that it opens beside the vagina, without the interposition of the normal strip of skin. Normal pregnancy and delivery is possible with this deformity.

The Vaginal Ectopic Anus. In this condition the anus is shifted further forward than in the preceding case, so that it opens into the lower part of the vagina. This is invariably described at present as 'imperforate anus with recto-vaginal fistula', a description which leads to a great deal of bad treatment. The reason is that a surgeon who looks at it in this way naturally tries to perforate into the bowel at the site of the normal opening, and ignores or tries to close the 'fistula'. The result is a bad type of colostomy in an inconvenient place, and misery for the patient. Once it is realized that the opening into the vagina is a true anus, complete with nerve and muscle sphincter mechanism, though misplaced and usually stenotic, treatment becomes much easier and more successful.

The principle of treating all these ectopic anuses is to get them to work where they lie with the least possible delay, the urgency coming from the tendency to develop colonic inertia which has been already mentioned. Once they are working properly, in the majority of cases nothing more need be done, and if a shifting backwards is necessary this difficult operation is much easier and safer several years later.

The treatment consists in a backward free incision, going well to the other side of the normal anal site, followed by prolonged dilatation. In the slighter degrees this will produce a shot-gun perineum, a condition which, as has been already stated, needs no treatment. If the opening is right inside the vaginal orifice, however, a transplantation backwards can be made, preferably at the age of 5 to 7 years. In doing this is is important to make a transverse incision and to leave a strip of intact skin between the new position and the vagina; the operation figured in many textbooks of a straight antero-posterior incision almost invariably results in the anus sliding forwards into the old position during the necessarily long and septic process of healing.

The Male Ectopic Anus. In this rare condition there is a stenotic opening well forward of the

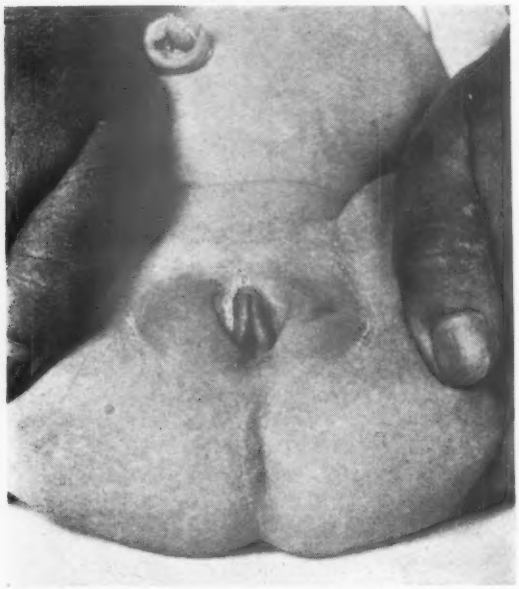

FIG. 3.—Covered anus and vagina. Excessive fusion of the genital folds has buried a normal vaginal opening and anal sphincter.

normal position, which tends to be ignored and an opening made in the normal position with the results already described. The treatment similarly consists in backward incision and dilatation; the final situation with a functionally normal anus lying slightly forward causes no inconvenience.

In both the male and the female varieties of ectopic anus which have had the stock erroneous treatment it is frequently sufficient to connect the stenotic congenital opening to the usually more stenotic surgically made one. It is most surprising how the sphincter mechanism will take charge of the anus thus produced, and most satisfactory to find that a simple cut with a scissors will transform a miserably dirty condition of the perineum into a functionally normal one.

The Covered Anus

The Covered Female Anus. In this condition the vulval opening is unduly short from before backwards, and there is no anal opening visible, though faeces are freely passed through the single orifice. A free backward incision will lay open a normally functioning anus, and if dilatation is persisted with new flexible skin will cover in the large raw areas surrounding this.

The Covered Male Anus. There are two main varieties of this malformation. In the first there is no anal orifice to be seen, but a small blue line can be seen running forward from the anal dimple for a variable distance. The blueness is due to the oozing of meconium along a sinus running just under the skin, and this sinus, produced presumably by undue fusion of the genital folds, may extend through the scrotum even well up the penis. The treatment is to open up the sinus and follow it back to where a normal anus can be exposed by a backward cut.

The second type is combined with a hypospadias cleft of the urethra, or, in one case of mine, with a large congenital fistula of the urethra. Here again the treatment is a free backward incision, which leaves a urethral opening on the very edge of the anus. I have found no difficulty, in the four cases in my experience, in constructing a new urethral tube to the end of the penis in one operation, using the buried strip method (Browne, 1949).

Deformities of the Rectum

Deformities of the rectum may be briefly mentioned. They mainly consist in a complete failure to form, often combined with a true fistula. In the male the fistula is most commonly into the prostatic part of the urethra, in the female into the upper part of the vagina; in neither case is there any sphincteric mechanism. As to treatment, this is an unsolved, and in many cases I am afraid, an insoluble problem. The one method I would give a strong warning against is the classical dissection upwards through the perineum. There are occasional successes recorded from this operation, but as those who record them do not recognize the microscopic nor the covered anus, I tend to put them down to these conditions. I personally have never met the condition of a simple diaphragm

FIG. 4.—Treatment of a similar case to Fig. 3. This shows the free incision necessary to expose the normal openings. If kept open by packing until the raw surfaces epithelialize, a process which occurs readily in this region, when a functionally normal perineum is produced.

across the anal opening which merely needs incision; and though this does not prove this condition does not exist, it certainly proves it to be rare.

The trouble with the dissection up through the perineum is that in the newborn infant this region is so small that it is impossible to deepen the incision to any extent without wrecking the whole pelvic floor. My present opinion is that the primary treatment of cases in which the rectum, as distinct from the anus, is malformed should be a colostomy of the transverse colon. This allows free access to the pelvis later on, which the almost invariable iliac colostomy does not.

A primary 'pull through' operation on the newborn encounters the difficulty that the blind end of the bowel is thin, grossly dilated, and full of sticky meconium. Later on the freeing of this blind end, and the pulling of it down through a hole stretched (not cut) in the pelvic floor holds some hope of success. It is certainly worth trying before condemning the patient to the only alternative, a permanent colostomy.

BIBLIOGRAPHY

Browne, Denis (1936). *Proc. roy. Soc. Med.*, **29**, 1409.
—— (1938). *Brit. med. J.*, **2**, 168.
—— (1949). *Postgrad. med. J.*, **25**, 367.
Russell, R. Hamilton (1922). *Brit. J. Surg.*, **9**, 502.
Swenson, O. and Bill, A. H. (1948). *Surgery*, **24**, 212.

THE CARDIO-OESOPHAGEAL SYNDROME IN CHILDHOOD

BY

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The term 'cardio-oesophageal syndrome' is used to cover all infants and children with incompetence of the cardia. Ninety-three instances of the syndrome have come to our notice during the last three and a half years. In some of these children the cardia was found to be below the diaphragm; in some, above the diaphragm (Table 1).

TABLE 1

CARDIO-OESOPHAGEAL SYNDROME IN 93 CASES

Sliding Hernia Cardia above Diaphragm (35 Cases = 39·5%)		Lax Oesophagus Cardia below Diaphragm (58 Cases = 69%)
35 + 4 = 39 Cases (42%)	←	4 Cases, Cardia herniated through Diaphragm
65%	Vomited from birth 6%	69%
93%	Vomited before 8 weeks 96%	100%
15%	Vomited projectile 25%	43%
51%	Haematemesis 37%	26%
52%	Gross loss of weight 36%	23%
5%	Visible gastric peristalsis 13%	19%
5%	Rammstedt's operation 10%	14%
100%	Cardia incompetent 100%	100%
Usually slightly dilated	← Oesophagus →	Often grossly dilated
High incidence	← Oesophagitis →	Low incidence
19%	← Definite ulcers →	None
19%	← Stricture →	None

It is proposed to divide the cases on the basis of the position of the cardia. There is a superficial similarity between the two groups but the course of the disease, the prognosis, and perhaps the aetiology, are not the same. We believe that the treatment is entirely different.

* A paper given at the inaugural meeting of the British Association of Paediatric Surgeons in London in July, 1954.

Group 1: Lax Oesophagus

When the incompetent cardia is situated below the diaphragm, we term the condition 'lax oesophagus'. It was first recognized as an entity distinct from hiatal hernia by Neuhauser and Berenberg in 1947. Although it is quite a common condition in babies, it has found scant recognition in the surgical literature (Berenberg and Neuhauser, 1950a and b). Neuhauser used the name 'chylasia of the cardia' because of the obvious incompetence of the cardiac orifice, but we prefer the name 'lax oesophagus', a term coined by our colleague, Dr. Saul Keidan (1953), because we think that it is not only the cardia which is relaxed but also the whole lower two-thirds of the oesophagus.

Incidence. It is difficult to form a definite idea of the incidence of the condition. The 58 cases included in this series were not all directly under our care; records were made available to us by the paediatricians working at the three children's hospitals on Merseyside. Mild examples of the syndrome treated as out-patients are not included in this survey. We compute that there are about 30 to 40 cases amongst the 41,000 babies born each year in the Liverpool region.

Sex. Of the 58 cases, 32 were boys and 26 were girls. There was one pair of fraternal twins, a boy and a girl. Apart from these twins, we have not had two cases in one family, but mothers have mentioned that previous children died or recovered from similar symptoms.

Birth Weight. The birth weights of our cases covered the normal range; few were much below average in weight.

Symptoms and Signs. Vomiting or regurgitation, or both, was the cardinal first symptom in every case. Vomiting dated from birth in a high proportion and almost invariably started before 2 weeks of age. The vomiting occurred during or between feeds and was projectile in 43% of cases.

The following points were noticed about the vomitus:

HAEMATEMESIS. Bright red or brown altered blood was visible to the naked eye in 26%. It was significantly less frequent than in the hiatal hernia group, due to a lower incidence of reflux oesophagitis.

BILE. A few babies vomited bile-stained material for a few days after birth.

MUCUS. Mucus was regurgitated or mixed with the vomitus from birth in some cases, but less often than in babies with hiatal hernia.

Gross loss of weight occurred in 23%. It was much less common than in the other group. This is, at least in part, accounted for by a more ready response to treatment. Failure to gain for longer or shorter periods was common.

Severe dehydration demanded parenteral fluids in 15%.

Anaemia due to bleeding or to malnutrition was found to some degree in about 40%.

Pain, discomfort, flatulence and general misery were noted in various degrees and combinations.

Visible gastric peristalsis was noted in 19% of cases. Visible peristalsis and an association with hypertrophic pyloric stenosis occurred more often in this group (Fig. 1). As our numbers are small, this is possibly not significant. It will be noticed that visible gastric peristalsis occurred more frequently than can be accounted for by the high incidence of pyloric stenosis; the same is true of projectile vomiting. Eight cases underwent Rammstedt's operation. Six of these operations were done while the babies were under our care; four were irrefutable cases of hypertrophic pyloric stenosis. Two babies were described as having a very small tumour, or some thickening of the pyloric region. Two cases were operated upon before coming to us and we have no certain knowledge of the condition of the pylorus.

FIG. 1.—Barium swallow showing typical picture of pyloric stenosis. There is free regurgitation from the stomach into the oesophagus. Pyloric stenosis with lax oesophagus.

FIG. 2.—Typical lax oesophagus. There is free regurgitation from the stomach into the dilated oesophagus.

It might be argued that in these eight children the cardiac reflux was due to back-pressure from the pyloric obstruction. The following points refute this suggestion: The uncomplicated case of pyloric stenosis never shows a patulous cardia on screening; these eight babies all continued to vomit and regurgitate after Rammstedt's operation, the adequacy of which was proved radiologically; visible gastric peristalsis was seen in a number of babies in whom no pyloric tumour was felt and in whom no radiological evidence of hold-up at the pylorus was demonstrated.

Radiological examination is required to prove

that an incompetent cardia is present and to decide its situation. The technique of the investigation is not easy and misleading reports are common unless the radiologist is used to dealing with young infants and is familiar with fluoroscopic appearances of the cardio-oesophageal region during swallowing in normal babies. The points to note are: (1) Free regurgitation from the stomach of the opaque material (Fig. 2); reflux is increased during inspiration, crying and coughing, in the Trendelenburg position, and if pressure is exerted over the abdomen, that is, when intra-thoracic pressure is reduced and intra-abdominal pressure is increased. (2) The cardia does not close but remains widely patent. (3) The normal acute angle at which the oesophagus joins the stomach on the left side is obtuse and may be equal on the right and left. (4) Muscular waves in the oesophagus are irregular and of small volume. (5) The oesophagus appears wider than normal and is often extremely distensible (Fig. 3).

FIG. 3.—Lax oesophagus showing enormous dilatation of the lower two-thirds of the oesophagus in a baby aged 3 days.

The very gross dilatation of the oesophagus in these cases is very remarkable; it is often present within a few days of birth, and sufficient to merit the term 'mega-oesophagus' (Fig. 3.) Only the lower two-thirds of the gullet is affected, that is, the plain muscle portion supplied directly by the vagus and which, according to Lendrum (1937), is supplied by the vagus only.

At oesophagoscopy the lumen of the oesophagus appears voluminous; gastric contents well up from the stomach and the instrument falls through the patulous cardia. The oesophago-gastric junction can be recognized without difficulty in most cases. Oesophagitis in various stages of acuteness and affecting various lengths of the lower third of the oesophagus may be seen. We have not seen a chronic peptic ulcer or stricture in a case in which the cardia remained below the diaphragm.

It has been suggested that the condition we call 'lax oesophagus' is in fact due to a small hiatal hernia which could be shown by careful screening. There is no doubt, however, that apart from four exceptions to be mentioned later, the cardia in these 58 children was below the diaphragm and remained there. This was proved by repeated radiography and oesophagoscopy.

Necropsy on three cases gave the following information:

The cardia, taken as the junction of stratified and columnar epithelium, was below the diaphragm. The cardia could not be pulled up into the chest until the membranes attaching the oesophago-gastric tube to the diaphragm had been divided. The hiatus was not considered to be unduly large, but it is difficult to assess the size of the oesophageal hiatus at operation, and probably more difficult at necropsy.

It is easy to understand that these children regurgitate and that regurgitation will be encouraged by recumbency and any factor which increases intra-abdominal pressure. It is not so easy to explain why vomiting, and more especially projectile vomiting, occurs. It is possible that the explanation lies in an associated hypertonicity of the alimentary tract below the cardia. This assumption would also explain (*a*) the high incidence of pyloric stenosis; (*b*) the occurrence of well-marked gastric peristalsis in cases in which there is no hypertrophy of the pylorus; (*c*) the bile-stained vomit seen in some cases soon after birth; (*d*) dilatation of the oesophagus in the absence of obstruction of the cardia; (*e*) the very interesting fact that one of our 'lax oesophagus' babies developed obstructive jaundice, which, at operation, was found to be due to spasm of the sphincter of Oddi.

Course of Disease, Prognosis and Treatment

The treatment of these cases is more or less standardized. They are nursed in the upright sitting position by means of pillows, harness, or by placing them in a padded box, to reduce regurgitation and make vomiting less easy. The feeds are thickened by the addition of 'nestergel', 'bengers' or cornflour.

Small, frequent feeds may be used so that the

stomach is not distended. Aludrox, or some other antacid, may be used to discourage oesophagitis. Anti-spasmodic drugs have little effect, though there has been an occasional apparent improvement with pylostropin.

The response to these measures is often dramatic; vomiting ceases, the child gains weight, and as the regime is gradually relaxed, symptoms do not recur. In some cases, improvement is slow or delayed, or symptoms recur as soon as the child is allowed out of the upright position. Many of these infants have been critically ill for weeks or months and have suffered serious complications (Table 2).

Four children with a proved lax oesophagus and a cardia below the diaphragm developed severe oesophagitis, and, over the course of months, the cardia was gradually pulled up into the chest. Three of these children later developed chronic oesophageal ulcer with stricture (Figs. 4, 5, 6 and 7).

TABLE 2

RESULTS IN 58 CASES OF LAX OESOPHAGUS

Medical Treatment

Satisfactory

Children doing well, vomiting has ceased, weight increasing .. .. 45 cases (79%)

Some had long periods in hospital or repeated admissions due to relapse; the following complications were recorded:

Gastro-enteritis ..	6 cases
Pneumonia ..	4 cases
Bronchitis ..	1 case
Lung abscess ..	1 case

Failed .. .. 13 cases (22·4%)

(*a*) Unsatisfactory. Child 1½ years old, still vomiting and much below expected weight .. .. .. 1 case

(*b*) Developed sliding hernia, and included under this heading .. .. 4 cases

(*c*) Operation, aged 5 months, repeated relapses and failure to gain weight .. .. 1 case

(*d*) Deaths due to the following causes .. .. 7 cases (12%)

- Gastro-enteritis
- Necrosis of lower end of oesophagus
- Oesophagitis and septicaemia
- Haematemesis
- Gastro-enteritis and lung abscess
- Inhalation of vomit
- Induction of anaesthesia for oesophagoscopy; poor-risk infant

(All cases were initially treated medically).

One child, still being treated medically at the age of 18 months, is still vomiting and regurgitating and much below weight.

Forty-five of the 58 cases (79%) are considered to have made, or to be making, a satisfactory recovery. Some of them have been radiologically examined after symptoms have ceased for various lengths of time and in these cases it has been demonstrated that the cardia has contracted, that reflux no longer occurs, and that the lumen of the oesophagus is narrowing.

One child, after repeated relapses, was operated upon at the age of 5 months and responded well to simple narrowing of the hiatus by stitches placed through the fibres of the right crus behind the oesophagus. At this single operation for 'lax oesophagus' the cardia was below the diaphragm. The hiatus did not appear larger than could be accounted for by the size of the oesophagus it accommodated.

Seven children died and the immediate cause of death is shown in Table 2.

To summarize, we think, in common with others, that the condition probably has a nervous or neuromuscular basis which involves the lower two-thirds of the oesophagus, the cardia, and perhaps the gastro-intestinal tract below the cardia. We realize that this is no real explanation. We are in doubt about the responsibility which the hiatus bears. The condition is often self-limiting and at least three-quarters of these children will recover with simple treatment.

Lax oesophagus cannot, however, be regarded with complacency as an unimportant condition out of which the child will grow. In our cases, serious complications were common. Many patients were in hospital for long periods, up to nine months, or had more than one admission. During this time they occupied badly needed medical beds and absorbed valuable skilled nursing time. From the mother's point of view, it cannot be much satisfaction to have a baby who sits in a box in a hospital ward. There was a mortality of 12%. We have no evidence that the course of the disease can or should be shortened by operation. If surgery were to be seriously considered, it would be difficult to decide its indications; during a bad phase, these babies would be poor risks, and when they improve, one cannot judge if the improvement is temporary or permanent.

Careful follow-up of these children and repeated radiological examination of those in whom vomiting persists, or in whom weight gain is unsatisfactory and those who relapse is indicated, so that the occasional case in which the cardia herniates above the diaphragm may be picked up as early as possible.

Group II: Sliding Hiatal Hernia

When the incompetent cardia is situated above the diaphragm the condition is usually called a sliding hiatal hernia. Findlay and Kelly (1931) and Kelly (1939) recognized that stricture of the oesophagus in children was associated with a partial thoracic stomach. Wyllie and Field (1946) described six children with partial 'short oesophagus and thoracic stomach' but did not mention that the

FIG. 4.—A series of barium meal swallows on the same patients taken at two-monthly intervals. This picture shows a typical lax oesophagus with free regurgitation.

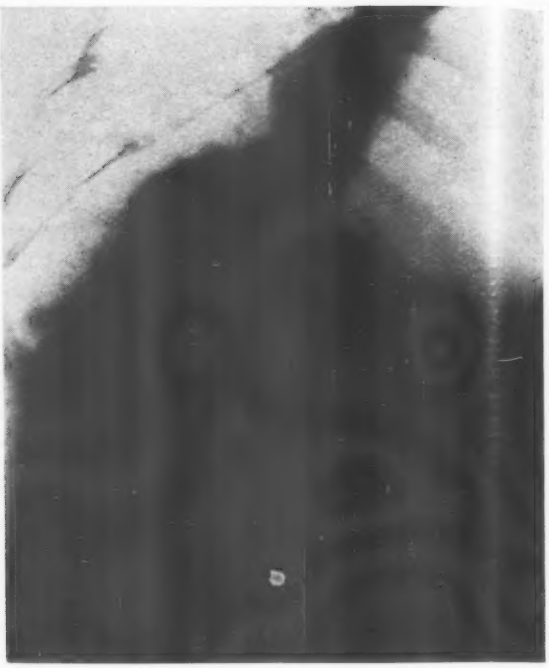

FIG. 5.—A series of barium meal swallows on the same patient taken at two-monthly intervals. In this picture the patient has developed a small hiatus hernia.

FIG. 7.—A series of barium meal swallows on the same patient taken at two-monthly intervals. In this picture there is an enormous hiatus hernia.

FIG. 6.—A series of barium meal swallows on the same patient taken at two-monthly intervals. In this picture the hiatus hernia has increased slightly in extent. There is a suggestion of stricture formation.

same symptoms are more commonly produced by an incompetent cardia below the diaphragm. There have been many excellent articles in the surgical journals on hiatal hernia during the last few years (Barrett, 1950; Allison, 1946, 1948, 1951; Harrington 1942, 1945; Belsey, 1954, etc.), but few surgeons have published a series of the abnormality in infants and children.

FIG. 8.—Barium swallow on a baby 48 hours old showing definite hiatus hernia.

In 35 of our 93 cases the cardia was above the diaphragm at the initial radiological examination. Some children were radiographed before the age of 4 or 5 days (Fig. 8), but most of them were a few weeks old and some older still.

At operation, the fibres of the right crus which form the hiatus invariably appear poorly developed, especially the part of the crus which forms the left and posterior aspect of the hiatus. In some cases, the posterior fibres are virtually absent. We think also that the diaphragm as a whole is often unduly thin, almost flaccid to the touch. We do not consider that the oesophagus is initially short; by and large, the younger the child the more easily can the cardia be placed below the diaphragm. Six cases were observed in which, during screening, the cardia was seen to be sometimes above and sometimes below the diaphragm; in all six cases it ultimately settled above. The four cases which started as lax oesophagus and went on to become sliding herniae are also significant. We have failed to find a description of a hernia of this kind in a stillborn infant. We do not certainly know the position of the cardia at birth. We do know that in at least some cases, the cardia has, at some time, been below the diaphragm and that it is drawn up through the hiatus. The migration of the cardia is probably dependent upon the difference in intra-thoracic and intra-abdominal pressures. We are of the opinion that a sac is present though it may be very small.

These facts suggest that the condition is probably a true herniation of the cardia from the abdomen into the posterior mediastinum. They do not prove that the condition is invariably produced in this way. Aetiologically, therefore, we consider that a poorly developed hiatus bears a heavy responsibility and is the 'congenital' element in this type of hernia. We consider that shortening of the oesophagus is secondary and results from oesophagitis and spasm, as in adults. In children, failure of the oesophagus to grow would ultimately produce relative shortening, and it is at least possible that this may be a significant factor in some cases. It will be seen that we do not consider the term 'congenital short oesophagus' justified.

Radiologically the oesophagus is not so dilated as in the lax oesophagus group. The diameter of the gullet is often about the same as that of the small segment of thoracic stomach. The supra-diaphragmatic stomach can be recognized radiologically by coarse vertical gastric rugae outlined by barium and a slight constriction at the cardio-oesophageal junction. On oesophagoscopy the junction can be identified by the transition from relatively smooth to coarse vertically ridged mucosa. The diaphragmatic pinch-cock is absent. Reflux is marked and constant. Oesophagitis is almost invariably seen; the degree of acuteness varies and it has a tendency to wax and wane.

The symptoms of hiatal hernia in infancy are due to the incompetence of the cardia and not to the cardia being situated above the diaphragm. We have, in common with others, had infants with

hiatal hernia in whom half or more of the stomach is above the diaphragm, and yet the cardia is competent, in spite of being divorced from the hiatus. In these cases symptoms may be entirely absent; if present, they conform to an entirely different pattern from the symptoms of the cases under discussion. The symptoms of these 35 children showed significant percentage differences from the lax oesophagus group. Projectile vomiting was less common; it is difficult to account for this. If projectile vomiting is a reflex phenomenon, one would expect it to be more likely to occur in the hernia group. Haematemesis occurred much more commonly due to the higher incidence of reflux oesophagitis. It is not clear why reflux oesophagitis is commoner in the hernia group as the same free reflux occurs in both. Gross loss of weight again was more often noted, at least in part, due to a less ready response to treatment. Visible gastric peristalsis was not seen, apart from the two cases of pyloric stenosis. These cases do not give the same evidence of being associated with a widespread neuromuscular imbalance as do children in the lax oeosphagus group.

Course of the Disease. In our experience, the response to posture is often unsatisfactory and a tendency to relapse marked. Complications fall into two groups: (1) The same tendency to contract chest infections, gastro-enteritis and to aspiration of vomit as the lax oesophagus cases; (2) complications in the oesophagus itself as follows:

SHORTENED OESOPHAGUS. Shortening of the oesophagus is a result of continued severe oesophagitis. It is known that once inflammatory changes brought about by the reflux of gastric secretions penetrate deep to the mucosa, fibrosis spreads up and down the oesophagus and results in shortening. It is also known that the different layers of the oesophagus are more loosely adherent in children, and that in them inflammatory spread and fibrosis occur more readily. Shortening of the oesophagus is common and may occur with astonishing rapidity.

PEPTIC ULCERS. In 30% of our cases there was radiological evidence of ulcer formation. The radiological findings were confirmed at oesophagoscopy. The ulcer was invariably in the oesophagus and usually within 1 in. of the cardia. We have not seen a chronic peptic ulcer in the thoracic portion of the stomach, either on screening or on oesophagoscopy. This important and interesting difference from sliding hiatal hernia of adults is probably related to the rarity of chronic peptic ulceration of the stomach and duodenum in children. We have not seen an ulcer at the cardia or in its immediate neighbourhood, and presume that the extreme lower end of the oesophagus is immune to the action of acid pepsin. One ulcer perforated into the mediastinum and at operation gave much the same picture as a chronic duodenal ulcer which has perforated into the pancreas.

STRICTURE. Some narrowing of the oesophagus occurred in 30% of cases. It was more common in cases with ulcer, but also occurred in cases in which a definite ulcer was not seen. The length of the narrowing varied from $\frac{1}{2}$ to $2\frac{1}{2}$ in.; it is difficult to assess radiologically as the barium is held up at the upper limit of the stricture and then trickles through in a thin stream, giving the impression of a long, narrow area. In the presence of spasm or stricture the proximal oesophagus dilates and hypertrophies (Figs. 9 and 10). In many cases, oesophagoscopy showed less narrowing than the radiograph had suggested; presumably spasm plays a part in producing the radiological appearances. Only one stricture required a gastrostomy.

Dysphagia due to a stricture can be temporarily relieved by dilatation. The amount of dilatation that can be obtained through an oesophagoscope in a small child is not very great. Dilatation encourages further reflux and perhaps encourages shortening, but as a temporary measure to improve swallowing and nutrition it has proved valuable.

With continued severe oesophagitis and associated spasm, or with the development of true

FIG. 9.—Barium swallow in a case of hiatus hernia with stricture showing marked dilatation of the proximal oesophageal segment.

narrowing, the symptoms change; solids and semi-solids are taken with more difficulty than liquids. Some mothers have stated that their baby is never happy until he has vomited. Older children complain of epigastric or retrosternal pain, discomfort or a sense of fullness after food. Some children become resistant to all nourishment.

Regurgitation is more common than true vomiting; mucus in large quantities is sometimes brought up after and between feeds and it may be blood-stained. Sometimes, the chief complaint is of the long time taken by the child to eat a small meal. Occasionally, children induce vomiting by putting their fingers down their throats. Weight gain is slow, or there is none; the patients are small for their age and tend to become self-centred and introspective. One or two children have given the impression that their only pleasure in life is to return, as soon as possible, any nourishment forced upon them.

Not all cases follow this severe course. A few improve with weaning, some have gained weight and appear well and happy with the cardia well above the diaphragm.

FIG. 10.—Same case as Fig. 9 after operation. The stomach is now well below the diaphragm. There is still slight dilatation of the proximal oesophagus. The level of the diaphragm is indicated by the two Michel's clips.

Operative Findings. The cardia has been found anywhere from just above the diaphragm to the level of D.6. (In one case which we had the opportunity of examining but not of operating upon, it was at the level of D.4-5.)

I consider that there is usually little difficulty in recognizing the position of the cardia at operation; there is generally a slight constriction at the cardio-oesophageal junction and a slight but definite change in the arrangement of the muscle fibres, and, I think, in the direction in which the arteries run. The level at which the cardia has been found at operation has usually conformed to radiological and endoscopic findings. In cases which have had prolonged oesophagitis, ulcers or strictures, there has been a varying amount of fixation of the oesophagus and it is firm and rigid to the touch. Enlarged lymph nodes are common, especially with chronic peptic ulcer.

In small children the vagi are usually of sufficient length not to form an obstacle to replacement of the cardia below the diaphragm. In cases where the cardia is high, and in older children, the vagi have sometimes appeared shorter than the freed oesophagus; twice we have divided the branches connecting the right vagus with the pulmonary plexus in order to get sufficient length.

The four cases of lax oesophagus which developed sliding hernia showed no variation from those in which the cardia was above the diaphragm when they were first examined.

As has been stated in speaking of the aetiology of the condition, the hiatus has invariably been found to be large and lax and to be, to some extent, developmentally deficient. The so-called oesophago-phrenic ligament appears to be a very definite structure; it is firmly attached to the gastro-oesophageal tube and has to be cut with scissors before the cardia can be brought down below the diaphragm. The small arteries to the oesophagus from the aorta appear to me to run transversely from aorta to oesophagus, not more and more obliquely upwards as the cardia rises, as one would expect.

Treatment. We are firmly of the opinion that sliding hernia in children is a surgical condition. It has been suggested that many cases will ultimately do well with medical treatment and that symptoms tend to subside at the time of weaning or later in childhood. The result of medical treatment, as exemplified by nine Liverpool cases, does not suggest that this is so (Table 3).

A child who continues to vomit and fails to gain weight is more likely to fall a victim to infection than is his healthy brother. There can surely be

TABLE 3
RESULTS IN NINE CASES OF SLIDING HIATUS HERNIA

Medical Treatment .. .. .. total number of cases	9
(*a*) Satisfactory; gaining weight, no vomiting	1
(*b*) Unsatisfactory; below weight for age, vomiting, etc. ..	5
Included are a boy of 13 years weighing 65 lb. who cannot eat solids, cardia at level of 5 T. stricture, and a boy of 7 years weighing 47 lb., repeated dilatation for stricture. The ages of the other children are 13 yr., 9 yr., 2 yr., 7 months and 6 months.	
(*c*) Still under observation at 10 weeks old	1
(*d*) Deaths	2
One child died aged 1 month, pneumonia.	
,, ,, ,, ,, 3 months, aspiration of vomit and haematemesis.	

Examples of cases in which medical treatment failed and operation was requested:
(1) Small hernia at 4 months, large hernia, stricture and perforation at 4 years.
(2) Recurrent pneumonia, repeated haematemesis, failure to gain at 5 months.
(3) Weighed 27 lb. at 3½ years; vomiting, haematemesis.
(4) Increasing hernia; shortening of oesophagus at 3 years.

little reason to allow a child to remain undernourished, and perhaps in pain, if his abnormality can be corrected. There is no guarantee that a child with a sliding hernia who does well on medical treatment will continue to do so, or that symptoms will not develop in later life when they may be more serious and less easily cured by operation.

There is no tendency for the cardia to become competent; it is permanently separated from the diaphragm and the hiatus is in any case inefficient. The valvular mechanism of the oesophago-gastric angle is obliterated and cannot develop as the gullet grows shorter. We therefore advise operation in all cases in which the cardia is above the diaphragm.

Time of Operation. Continued vomiting and failure to gain weight, severe, persistent or recurrent oesophagitis, progressive shortening of the oesophagus, peptic ulcer of the oesophagus and narrowing of the oesophagus are all absolute indications for operation.

In a child who is symptom free or in whom symptoms are mild, there is no urgency and operation can be undertaken at any convenient time, provided the position of the cardia is checked at frequent intervals.

If operation is advised for all cases it must cure symptoms and prevent complications, give a low recurrence rate and be safe (Table 4).

The important points in successful operation are adequate freeing of the oesophagus up to or above the aortic arch and complete separation of the oesophago-gastric tube from the diaphragm; firm fixation of the fundus of the stomach to the undersurface of the left dome of the diaphragm; careful repair of the hiatus.

We have found a thoracic approach to the oesophagus and a transdiaphragmatic approach to the stomach satisfactory, and since using this method, we have, so far, had no recurrence in 27 cases. Previously we operated on seven cases

TABLE 4
RESULTS IN OPERATIONS FOR CASES OF SLIDING HIATUS HERNIA

Surgical Treatment .. .. .. total number of cases	30
(*a*) Operation above diaphragm	7
(i) Recurrence	4
(ii) Satisfactory	3
(*b*) Operation above diaphragm and through diaphragm	20 + 4 reoperated
(*c*) Operation above diaphragm and through diaphragm and dilatation of stricture through gastrostomy	3
(*b*) and (*c*)	27
Satisfactory; relief of symptoms and no recurrence of hernia to date	29
Total operations	33
Mortality	1

without opening the diaphragm and had four recurrences. These recurrent cases were later on successfully operated upon by the transpleural transdiaphragmatic approach. Severe strictures are dilated with Hagar's dilators through a small gastrotomy while the chest is open; the three cases so treated have not required post-operative dilatation. The children have stood operation well and post-operative complications have, on the whole, been few and transient. All cases are free of symptoms, except that two children have been slow to gain weight and appetite, in spite of normal or near normal appearances on radiography and oesophagoscopy. The longest follow-up in this small series, is, however, only three and a half years.

To conclude, we believe there is some evidence that lax oesophagus and sliding hiatal hernia are aetiologically distinct conditions. Further study of the former is required to settle the nature of the responsible factor. In sliding hiatal hernia we do not know whether the cardia is ever above the diaphragm at birth. It is natural to want to know the answers to these fascinating problems, but the burden of this discourse is the importance of the position of the incompetent cardia. I reiterate words written by Allison in 1948 about sliding hernia in children:

> 'The condition is of particular interest to paediatricians because its early recognition may lead to surgical reduction of the hernia, and for the patient, escape from a life of dysphagia and malnutrition.'

I wish to acknowledge the assistance of Mr. P. P. Rickham who performed about half the operations, most of the endoscopic examinations and a large proportion of the time-consuming radiological examinations.

REFERENCES

Allison, P. R. (1946). *J. thorac. Surg.*, **15**, 308.
—— (1948). *Thorax*, **3**, 20.
—— (1951). *Surg. Gynec. Obstet.*, **92**, 419.
Barrett, G. M. (1950). *Brit. med. J.*, **1**, 245.
Belsey, R. (1954). *Ann. roy. Coll. Surg. Engl.*, **14**, 303.
Berenberg, W. and Neuhauser, E. B. D. (1950a). *Amer. J. Dis. Child.*, **79**, 402.
—— —— (1950b). *Pediatrics*, **5**, 414.
Findlay, L. and Kelly, A. B. (1931). *J. Laryng.*, **46**, 797.
Harrington, S. W. (1942). *Ann. Surg.*, **115**, 705.
—— —— (1945). *Ibid.*, **122**, 546.
Keidan, S. E. (1953). Lecture to the Liverpool Paediatric Club.
Kelly, A. B. (1939). *J. Laryng.*, **54**, 621.
Lendrum, F. C. (1937). *Arch. intern. Med.*, **59**, 474.
Neuhauser, E. B. D. and Berenberg, W. (1947). *Radiology*, **48**, 480.
Wyllie, W J. and Field, C. E. (1946). *Archives of Disease in Childhood*, **21**, 218.

TWO CASES OF OESOPHAGEAL STRICTURE IN CHILDHOOD REQUIRING RESECTION

BY

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(RECEIVED FOR PUBLICATION OCTOBER 18, 1954)

Reflux oesophagitis appears to affect principally two great groups of patients, infants on the one hand and patients of late middle age on the other. The intervening years seem to be comparatively free from the condition and it is relatively uncommon for symptoms to occur for the first time in late childhood or early adult life. An exception to this generalization is the patient who develops oesophagitis in association with excessive vomiting of pregnancy. Most of the affected infants recover from their early oesophagitis either spontaneously or as the result of treatment, a few die, and in the remainder symptoms of oesophagitis persist.

One of the patients described below suffered from persistent symptoms starting in infancy and in the other the condition appears to have shown itself for the first time at the age of 13 years. In both patients resection was carried out and neither child has had any further symptoms.

Case Reports

Case 1. K.H., a girl aged 14 years, was referred from the Hammersmith Chest Clinic on August 10, 1951, with the complaint of recurrent attacks of 'vomiting' after meals.

She had been a normal healthy baby, breast fed till 1 year old. Subsequently there had not been any illness of note until September, 1950, when the present complaint began. The 'vomiting' occurred immediately after taking food and was associated with a feeling of sticking in the midsternal region. Pain was absent. The remittent attacks were precipitated by solids and were associated on some occasions with difficulty in swallowing liquids.

In the two months before consultation her symptoms had increased in severity. The patient's weight was 86 lb. (39 kg.). Physical signs of note were lacking. A barium swallow showed an oesophageal stricture at the level of the eighth thoracic vertebra and an associated hiatus hernia (Fig. 1).

The child was admitted for treatment. A diet of fortified milk was given together with olive oil and alkaline powders. In bed, an upright posture was maintained day and night. Twice daily autodilatation was carried out with gum-elastic bougies.

The symptoms improved steadily and by August 27, 1951, solids could be swallowed without regurgitation. She was discharged to attend as an out-patient, while maintaining the above régime, with the substitution of a normal diet for the fortified milk.

FIG. 1.—Radiograph showing oesophageal stricture and hiatus hernia.

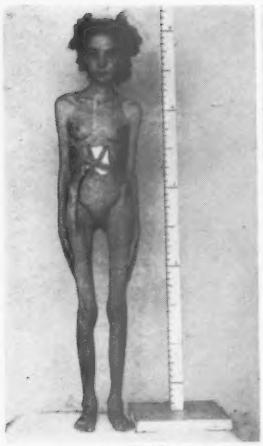

FIG. 2.—K.H. before resection of the stricture.

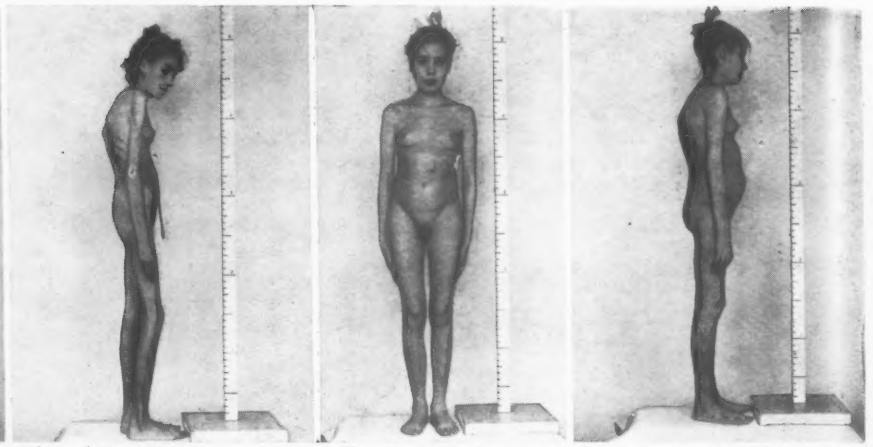

FIG. 3.—K.H. three months later after resection of the stricture.

Unfortunately the patient defaulted and treatment was erratic. By February 26, 1952, severe symptoms had recurred, bougies could not be passed, and her weight had fallen to 73 lb. (33·1 kg.). She was re-admitted and the original régime was re-instituted with a slow but steady improvement. On discharge on May 2, 1952, solids could be swallowed with ease and her weight had increased to 100 lb. (45·4 kg.).

After her discharge, however, she again relapsed and on readmission on September 20, 1952, dysphagia had been severe for four weeks. The patient's weight had fallen to 55 lb. (24·9 kg.). A follicular rash was present but was limited to the abdomen and legs. The blood urea level was 206 mg. % and haemoglobin 21·6 g. (147%).

Through a Ryle's tube an intragastric milk drip was started and simultaneously fluid was given per rectum. By the tenth day a positive fluid balance was being maintained, her weight had increased to 58 lb. (26·4 kg.) and the rash was fading. The blood urea had fallen to 48 mg. %. The Ryle's tube was removed and bouginage restarted. Unfortunately, following a period of continued improvement, a severe relapse recurred, her weight fell rapidly, and it proved impossible to re-insert the Ryle's tube.

On October 14, 1952, an inkwell type of gastrostomy was performed (Fig. 2). A month later, her weight was 67 lb. (30·4 kg.). It was still impossible to pass a bougie. Because of this and because out-patient treatment had proved unsatisfactory, resection was advised.

At operation (R.H.F.) on November 18, the chest was opened through an incision in the eighth intercostal space on the left side. The left pulmonary ligament was divided and a hiatal hernia was at once apparent. Halfway between the arch of the aorta and the diaphragm the oesophagus was thickened and enlarged for a distance of 1½ in. There was associated peri-oesophagitis. The resulting mass felt stony hard and the impression on palpation was that the condition was indistinguishable from a carcinoma, although in view of the circumstances it was thought that the correct diagnosis was a chronic ulcer. The incision was prolonged inwards to divide the costal cartilage and the diaphragm was opened. The spleen was excised and the stomach mobilized by dividing the gastrocolic omentum and the left gastric artery and vein. Another intercostal incision was made through the fourth space and the oesophagus was freed to within ¾ in. of the arch of the aorta. The stomach was divided obliquely so as to remove most of the lesser curvature. The distal cut end of the stomach was closed with a double row of sutures and the rest of the stomach and the lower part of the oesophagus were removed. A separate incision was made in the closed distal part of the stomach and this was anastomosed to the cut end of the oesophagus just below the arch of the aorta. The diaphragm was approximated around the stomach and the chest closed around a large intercostal drain. The right pleura was widely opened during the resection because of pleural adhesions and an intercostal drain was introduced on the right side at the end of the operation. Both drains were led into waterseal bottles.

The girl made a very satisfactory recovery and was

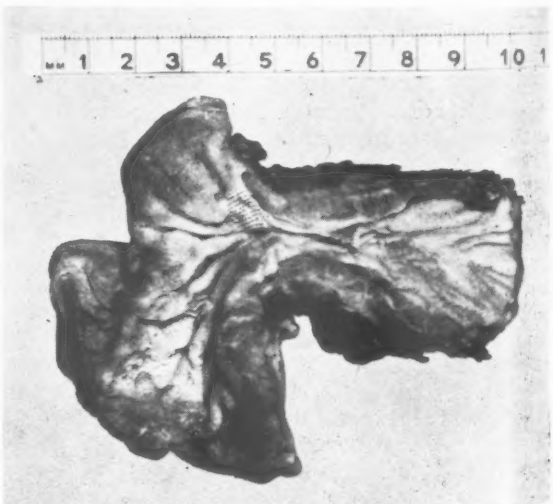

FIG. 4.—Part of the oesophagus resected showing the stricture.

FIG. 5.—Histological appearance of the stricture. Areas of osteoid tissue can be seen in the middle of the scar and the oesophageal mucosa shows ulceration.

discharged on January 15, 1953, her weight then being 101 lb. (45·8 kg.), and her clinical condition excellent (Fig. 3). She continued on an oesophagitis régime with alkalis and olive oil.

When last seen on May 18, 1954, her weight was 110½ lb. (50·1 kg.). For the previous five months she had not taken any medicine and had eaten a normal diet without any trouble.

The specimen (Fig. 4) showed that the stricture was caused by an excess of fibrous tissue involving all coats. In the midst of the scar tissue were areas of osteoid change (Fig. 5). In the oesophageal mucosa was a small ulcer with evidence suggestive of healing.

Case 2. C.T., a girl of 9, was referred for treatment by Mr. Alan Small.

After an uneventful first year of life, she started to have attacks of 'vomiting' with meals. Each attack lasted three or four days and recurred every two to four weeks. There were neither associated symptoms nor physical signs. A diagnosis of mental retardation was made and maintained until the summer of 1952. By then only liquids could be swallowed and her weight was falling rapidly. She was referred to Mr. Small, who made a diagnosis of oesophageal stricture and hiatus hernia.

On admission on September 5, 1952, her weight was 53½ lb. (24·3 kg.). The presenting symptom was effortless regurgitation on eating solids. A barium swallow confirmed the presence of a hiatus hernia with an oesophageal stricture at the level of the eighth thoracic vertebra (Fig. 6).

A régime similar to that described in Case 1 was started. On September 13, 1952, her weight was 62 lb. (28·1 kg.). Three days later oesophagoscopy showed a marked gastric reflux, but no inflammatory changes or obvious stricture. The previous treatment was continued but attempts were made to convert the liquid diet to a solid one. By October 28, 1952, it had proved impossible on several occasions to institute this change without the recurrence of vomiting. Having regard to the patient's age and symptoms together with the oesophagoscopic findings, it was decided to try the effect of a repair of the hernia combined with a Heller-type operation on the spasmodic stricture.

At operation (R.H.F.) on October 28, the chest was

FIG. 6.—Radiograph showing stricture of the oesophagus with hiatus hernia.

opened through an intercostal incision through the eighth left interspace. A hiatal hernia was present and the pleura overlying the oesophagus seemed to be very much thicker than normal and contained more blood vessels than usual. The left pulmonary ligament was divided to expose the lower part of the oesophagus and it was then seen that there was a stricture, ½ in. in length, 2½ in. above the level of the diaphragm. At the level of the stricture the pleural adhesions appeared to be particularly thick and the oesophagus itself was thicker. An incision was made in the diaphragm away from the crus and the presence of the hernia confirmed. There appeared to be about 1 in. of stomach in the thorax, but it was difficult to determine the exact junction of the oesophagus and stomach. The muscles over the stricture were divided longitudinally and the epithelium was inspected. It appeared to be scarred. The lower oesophagus and cardiac end of the stomach were mobilized and the stomach was replaced below the diaphragm and kept in position with interrupted thread sutures passing through the phrenico-oesophageal ligaments and the crus of the diaphragm. The diaphragm was closed and an intercostal waterseal drain introduced and the thoracic wound closed.

The child made a good recovery from the operation but the difficulty in swallowing persisted and it had to be recognized that the operation had been a failure. It was, therefore, decided to carry out a radical excision of the affected part of the oesophagus.

At operation (R.H.F.) on December 2, the chest was re-opened through the same incision. The diaphragm was opened and the incision carried right down to the

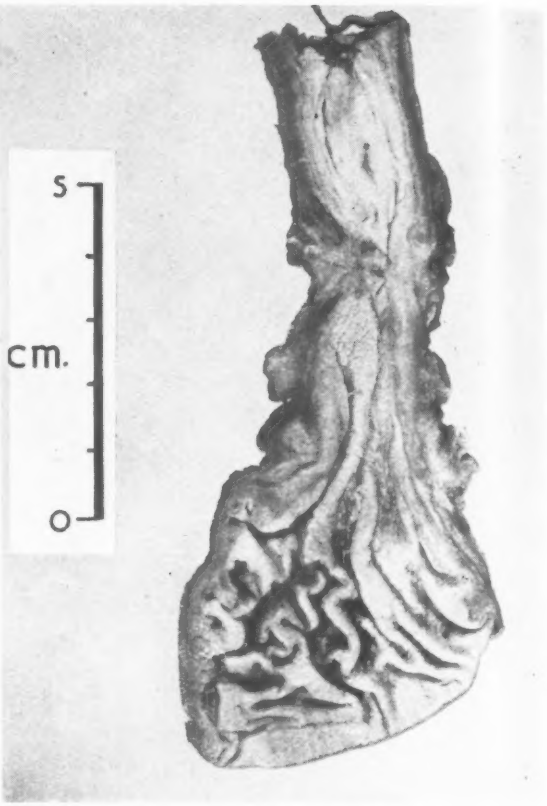

FIG. 8.—Operation specimen.

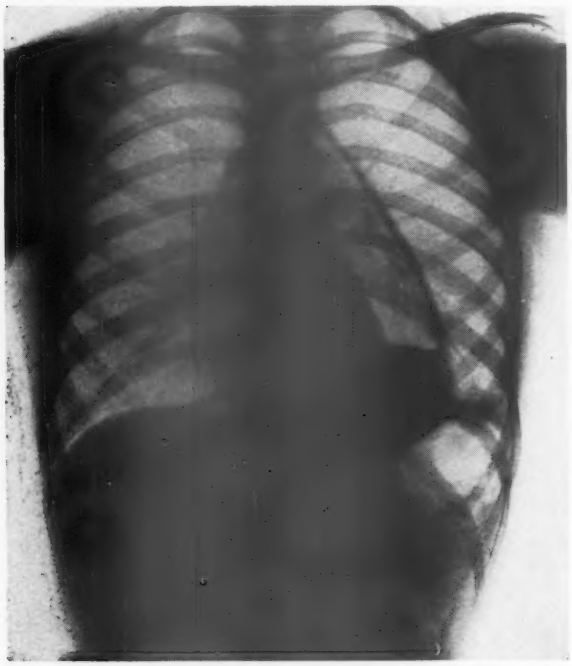

FIG. 7.—The oesophagus containing the stricture has been resected and the stomach anastomosed to the stump of the oesophagus above the arch of the aorta.

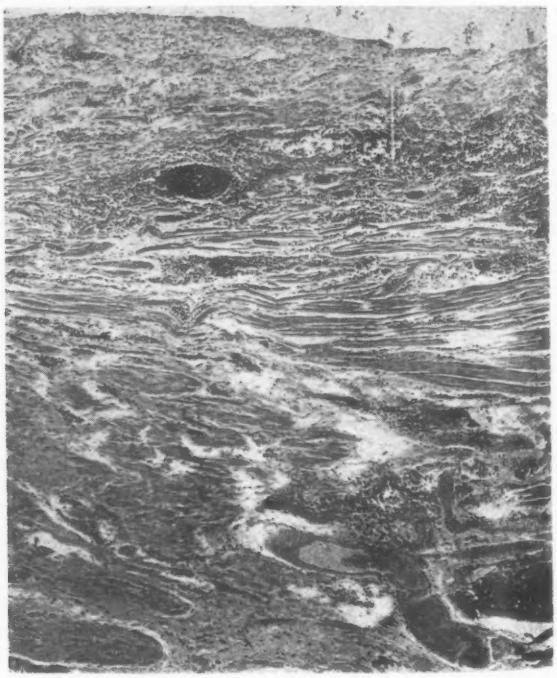

FIG. 9.—Histological appearance showing absence of epithelium of both cardia and oesophagus.

oesophagus, dividing the right crus. The spleen was removed and the stomach was mobilized by ligaturing and dividing the gastro-colic omentum and the left gastric vessels. The mobilization was carried up to the lower half of the oesophagus. To obtain a good view of the oesophagus above the arch of the aorta, a second intercostal incision was made in the fourth interspace after drawing the scapula well forward. The oesophagus above the arch of the aorta was exposed by dividing the overlying pleura and the oesophagus itself was divided at the level of the arch of the aorta. The stomach was divided obliquely so that the line of section was from the fundus down to the middle of the lesser curvature. The cut end of the stomach was sutured and the fundus brought up in front of the arch of the aorta and anastomosed to the cut end of the oesophagus after having divided the pyloric sphincter without opening the lumen. An intercostal waterseal drainage tube was introduced and the rest of the incision closed.

Recovery was uninterrupted and symptoms vanished. At Christmas, 1952, the patient consumed a large dinner with ease (Fig. 7). At discharge on January 12, 1953, her weight was 62 lb. (28·1 kg.).

By April 15, 1954, when she was admitted for excision of the tonsils and adenoids, the girl was asymptomatic nor was she keeping to an oesophagitis régime. Her weight was 65½ lb. (29·7 kg.).

The specimen (Fig. 8) shows the site of the stricture. Histologically the epithelium, of both cardia and oesophagus at their junction, is either debased or absent. Where it is absent the lumen is lined with organized granulation tissue (Fig. 9).

Summary

Two patients are described in whom severe stricture formation occurred in the oesophagus, presumably the result of reflux oesophagitis. The process was not affected by any conservative attempts at control and resection had to be undertaken.

The histological appearance in Case 1 showed areas of osteoid tissue which seems an unusual finding in strictures of this nature.

BIBLIOGRAPHY

Belsey, R. (1952). In *Modern Trends in Gastro-enterology*, Ch. 7, p. 128. Ed. Jones, F. A. London.

Kelly, A. B. (1930a). *Proc. roy. Soc. Med.*, **23**, 1521.

—— (1930b). *J. Laryng.*, **45**, 680.

Sheldon, W. and Ogilvie, A. G. (1929). *Archives of Disease in Childhood*, **4**, 347.

ACUTE DISTENSION OF THE GALL BLADDER IN CHILDREN

BY

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Diseases of the gall bladder in childhood are of great interest. Recently, Forshall and Rickham (1954) and previously Potter (1938) drew attention to cholecystitis associated with gall stones. The majority of such cases do not present as acute emergencies. Fèvre (1933) states that diseases of the biliary tract presenting as surgical emergencies are usually a perforation or volvulus of the gall bladder. There is, however, a third lesion which he has encountered, namely, acute distension of the gall bladder. This paper reports two such cases and supports views already held by French surgeons.

Case Reports

Case 1. A 4-year-old boy had been well until the day before admission. He then developed severe abdominal pain, intermittent in character, and suffered from nausea with occasional vomiting. The bowel movements had been normal but his urine was stated to have been dark. The pains lasted for some hours and then subsided, but soon returned and were apparently very severe though some relief was obtained by flexing the trunk. The pain was not referred to the gall bladder region but to the central abdomen. He was admitted to hospital with the tentative diagnosis of acute appendicitis.

He had complained of mild central abdominal pain for six months, but in general his health was good.

On admission the temperature was 99° F., pulse rate 132, and respiratory rate 24 per minute. The child lay quietly and was not in severe pain. The face was flushed, the throat injected, skin and sclerae were clear. There was marked tenderness of the right hypochondrium and epigastrium. Tenderness was elicited in the right iliac fossa but here there was no rigidity. It was possible to feel the edge of the liver but there was no local swelling nor was any mass palpable in the abdominal cavity. Bowel sounds were present in all areas. Rectal examination was negative. A rectal wash-out revealed pale faecal material, some mucus, and a fleck of red blood. Bile was not present in the urine.

Shortly afterwards the child had a severe attack of pain; he vomited profusely, and there was marked rigidity in the right hypochondrium. A tentative diagnosis of intussusception was made and the child prepared for operation.

Under anaesthesia a tense mass was palpable in the right hypochondrium. On opening the abdomen clear fluid escaped. Numerous enlarged, injected and firm lymph nodes were palpable in the mesentery and duodeno-jejunal flexure The caecum was located, the appendix was not inflamed but was removed. There was no abnormality in the small intestine and the large gut was traced uneventfully to the rectum. On displacing the transverse colon downwards, a large, tense gall bladder presented. Owing to extreme distention, it was not possible to determine the presence or absence of stones in the viscus but none were found in the cystic duct or common ducts neither of which was dilated. The gall bladder was essentially normal in colour; there was no thickening or obvious signs of infection. Further pressure did not empty it. Because of the distension packs were inserted, and the operative field was prepared as for cholecystectomy. As the organ was so normal apart from its distension, it was decided that aspiration would suffice and 40 ml. of dark green bile were aspirated. The needle was removed and the puncture sealed with a purse string suture. The abdomen was closed without drainage.

Apart from mild pyrexia for 48 hours convalescence was uneventful. When reviewed four months later the patient was well. Pain had not recurred.

Case 2. A 10-year-old boy had been well until four days before admission. He complained of severe abdominal pain, umbilical in distribution. His appetite had been poor, he had had some nausea, but had not vomited. As the pain subsided, he went to school the next morning, but was sent home in the afternoon owing to a recurrence of abdominal symptoms. At home his doctor noted increased tonicity of the right rectus muscle, particularly in its upper half. Tenderness was elicited over the distribution of the small bowel mesentery. The condition again subsided until the afternoon of the day of admission, when the child was nauseated, and severe pain had recurred. The right rectus muscle was in acute spasm and there was marked rigidity in the right hypochondrium. During transit to hospital by car, over a route paved with cobblestones, the child com-

plained bitterly of the pain, which was accentuated by the jolting.

Before his illness the child had been well, but his appetite had been variable and he was fussy about food, not liking eggs and fat. He had received treatment for a threadworm infestation.

On admission the temperature was 98° F., pulse rate 96 and respiratory rate 22 per minute. The child lay quietly in bed. The face was pale and the fauces were clear, as were the skin and sclerae. There was marked tenderness and rigidity in the right hypochondrium. The child could point to the site of maximum tenderness in the subhepatic region, 2½ in. above McBurney's point. He resented palpation in the area. He was seen by the senior surgeon who made a pre-operative diagnosis of inflammation of a high-placed appendix and cholecystitis. In view of the definite rigidity, which could not be ignored, the child was prepared for the theatre.

At operation by Mr. Matthew White, through a right rectus incision the appendix was located in the right iliac fossa. It was injected but not acutely inflamed and was removed. Numerous mesenteric nodes were present. The gall bladder was tensely distended, and it was obviously the cause of the abdominal tenderness. The gall bladder was normal in colour and appeared slightly thickened. The remarkable tension within the viscus prevented excluding the presence or absence of a calculus. The lymph nodes in the region of the cystic duct was palpable, firm and enlarged. Some doubt arose as to whether one was indeed a node or a stone impacted in the cystic duct. The common bile duct was not dilated and so consequently 30 ml. of bile was aspirated. The bile was green and contained mucus. A cholecystostomy was performed, no stones were found, and the abdomen was closed.

Section of the appendix showed a mild acute catarrhal reaction, and some tomato pips in its lumen. Culture of the bile was sterile.

For the first four post-operative days the child ran a temperature of 99 to 101° F. Seven ounces of bile were drained in the first 24 hours. On the fourth day the urine was dark but no bile was present. No icterus occurred but a transitory pruritis of the chest wall. The bowels moved on the fifth day and were normal in colour and contained bile pigment. Thereafter recovery was uneventful and the patient was discharged on the sixteenth post-operative day. One month later threadworms were found in a bowel wash-out.

Discussion

These two cases were acute surgical emergencies and were not associated with stone formation. It is emphasized that in such cases the gall bladder is not only tense but is grossly distended. In both cases 30 ml. of bile were aspirated and despite this the gall bladder was by no means emptied. Following aspiration the gall bladder was still larger than usual.

The pathology of the condition is obscure; a biliary dyskinesia could be postulated, but the condition appears to be the result of an inflammatory reaction of the cystic lymph node. In both cases there was an extensive and acute mesenteric adenitis. In the second case the cystic node was palpable. Due to unfamiliarity with this condition, an accurate assessment of the node was not made in Case 1. The cystic duct in both cases appeared to be normal but a larger lymph node could have obstructed the cystic duct and a situation might have arisen as described by Lee and Englender (1953) in which a hydrops of the gall bladder was found which actually caused a pyloric obstruction; this case had an ascaris infestation. In Case 2 a known threadworm infestation was being treated.

The pain in these cases is severe and can simulate a gall bladder colic. The child has relief for some time and then the pain recurs. The rigidity of the rectus muscle is marked and from its site and intensity may suggest an underlying perforation. The French writers have commented upon this feature. The pre-operative diagnosis is infection of a highly situated appendix or intussusception. Acute cholecystitis may be considered. Ombrédanne (1946) states bluntly that such cases are 'too easily diagnosed as appendicitis'. The cases presented similarly to those described by Fèvre (1933) and at operation almost identical findings were observed.

Discussion will always arise as to what operative procedure should be carried out when confronted with this condition. Such cases are usually met with by the younger surgeon. It is recommended that simple aspiration of the gall bladder will suffice. This procedure was effective in my first case, and was the method chosen by Fèvre in his two cases. Cholecystostomy should be reserved for those cases in which there is doubt as to whether the bile is infected or in which there is a possible associated cholelithiasis. The observations of Edington (1925, 1930) suggest that in more experienced hands the latter complication should be dealt with by cholecystectomy. Where gross pathological changes in the gall bladder and cystic duct are noted cholecystectomy is deemed advisable.

My findings in this condition are in accordance with and support the opinions of the French surgeons who drew attention to this interesting clinical entity.

My thanks are due to Mr. A. P. Laird, under whose care the patients were admitted, for permission to publish these cases, and to Mr. Matthew White whom I assisted in the second case.

References

Edington, G. H. (1925). *Brit. med. J.*, **1**, 1126.
—— (1930). *Ibid.*, **1**, 782.
Fèvre, M. (1933). *Chirurgie Infantile d'Urgence*, p. 310. Paris.
Forshall, I. and Rickham, P. P. (1954). *Brit. J. Surg.*, **42**, 161.
Lee, C. M. and Englender, G. S. (1953). *Paediatrics*, **11**, 449.
Ombrédanne, L. (1946). *Précis Clinique et Opératoire de Chirurgie Infantile*, 4th ed., p. 542. Paris.
Potter, A. H. (1938). *Surg. Gynec. Obstet.*, **66**, 604.

A SURGICAL AND BIOCHEMICAL STUDY OF A CASE OF BILATERAL HYDRONEPHROSIS AND HORSESHOE KIDNEY

BY

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Congenital abnormalities of the kidney are often symptomless and may be discovered on routine investigation of minor urinary disorders. Such kidneys are, however, very susceptible to pathological change and have been described in association with every form of renal disease. A horseshoe kidney is the commonest form of fused kidney and is often associated with hydronephrosis of one or both sides. The hydronephrosis is usually attributed to kinking of the ureter by bands or vessels and occasionally to compression of the ureter as it passes over the isthmus. The important pathological point is how much damage is done by the hydronephrosis to the renal tissue, as the nephrons can be damaged both by back pressure and by interference with their blood supply. The extent of the obstruction and superadded infection will have bearing on how great this damage is.

Such lesions may be asymptomatic or give rise to a variety of symptoms. The commonest are failure of growth, abdominal pain, and symptoms related to micturition. In many cases there is a palpable abdominal mass. If both halves of the kidney are functioning normally intravenous pyelography will give the diagnosis in almost every case. If hydronephrosis has suppressed function on one side a malrotation of the other side may be diagnosed. Retrograde pyelography should, however, provide the true answer.

The treatment, if possible, should consist of relieving obstruction and preserving the kidneys. In simple hydronephrosis the results are excellent, but in the case of a horse-shoe kidney with uni- or bi-lateral hydronephrosis the position is more difficult since probably neither side is functioning normally. It is therefore essential to find out as far as possible what function is present on either side. Surgery must be conservative in these cases, but can produce a great deal of benefit.

The case presented here showed several points of interest. The diagnosis was fairly simple but illustrates some of the points mentioned above. Biochemical studies of the urine produced by the two halves of the patient's kidney were puzzling but extremely interesting, and we will attempt to explain them. It is far too early to say what the child's future will be, but the results of surgical treatment are, at least during the first year afterwards, very impressive.

Case Report

The patient, a girl, was admitted to hospital at the age of 6 months, on November 9, 1953. Her mother complained that she failed to thrive. Her birth weight had been 6 lb. 2 oz., and at 10 days of age she had regained this weight. One week later she became 'grizzly' and started to lose weight. At 5 weeks she weighed 5 lb. 8 oz. and from then until the time of admission, when she weighed 10 lb., she had gained extremely slowly, often having periods of loss. She had vomited very occasionally, but had no other symptoms. She was breast fed for six weeks, and then was put on National dried milk. Adequate amounts had always been offered to her but she seldom took even half the feed.

Her mother was quite well during her full-term pregnancy and the birth was quite normal. She had a healthy brother of 5 years, and both parents were quite well. Her paternal grandfather and an uncle died of tuberculosis, but many years before her birth, and the father on several occasions had a negative chest radiograph.

On examination the baby weighed 10 lb. and was 23¼ in. long. Her temperature was 102°. Her blood pressures were: right arm 180/80, left arm 190/120, right leg 170/110, left leg 190/40 mm. Hg. There was a loud systolic murmur, maximal in the fourth left interspace. The abdomen was distended, and in the left hypochondrium and lower abdomen a large cystic mass could be felt. There were no other abnormal signs.

A blood count gave: red cells 3,620,000, haemoglobin 76% (10·6 g.), white cells 11,000 (63% neutrophils, 37% lymphocytes), E.S.R., 30 mm.

No albumin or pus cells were found in the urine which was sterile on culture.

A blood culture was negative.

The Mantoux test (1 : 100) was negative.

A chest radiograph was normal, and no abnormality was shown in a straight radiograph of the abdomen.

FIG. 1.—Intravenous pyelogram.

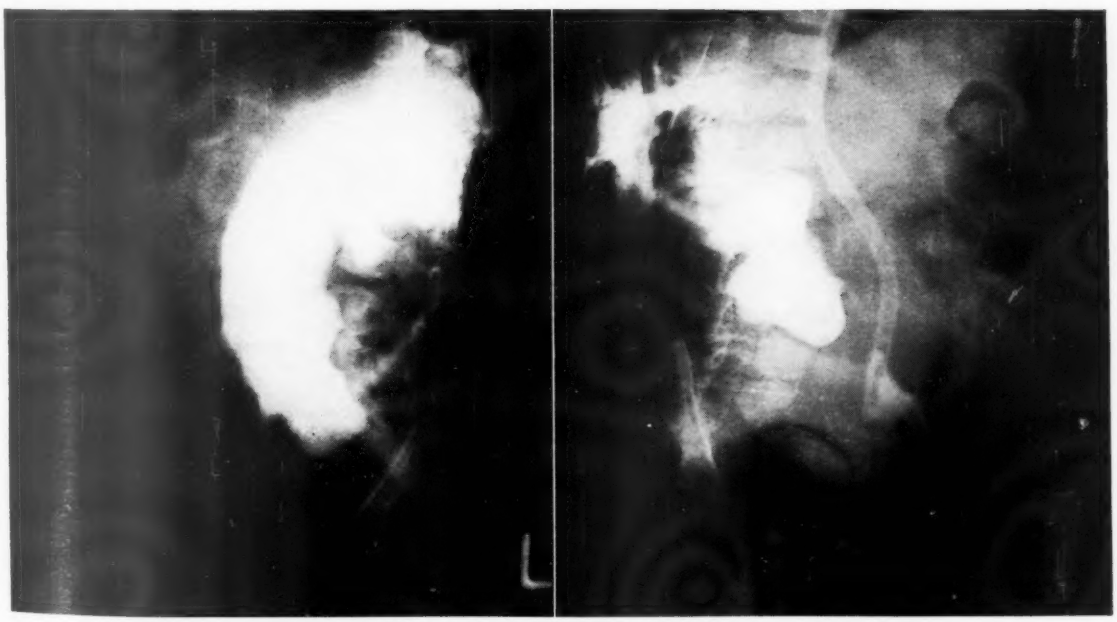
Blood analyses gave: Urea 84 mg. per 100 ml., phosphorus 3·3 mg. per 100 ml., calcium 10·6 mg. per 100 ml., bicarbonate 37·5 vol. per 100 ml., sodium 308 mg. per 100 ml., potassium 19·0 mg. per 100 ml., chlorides 611 mg. per 100 ml., and total proteins 6·71% (albumin 3·29%, globulin 3·42%).

An intravenous pyelogram showed bilateral hydronephrosis, more marked on the left side.

A series of chemotherapeutic and antibiotic drugs was tried with no effect on the patient's persistent temperature. Excessive hydration both orally and intravenously was similarly ineffectual. However, her general condition remained good, and surgical exploration was decided upon in spite of her fever. The intravenous pyelogram showed hydronephrosis without indicating the site of the obstruction (Fig. 1) but clinically the swellings were so low in the abdomen that it was thought that the ureters must be dilated. In view of the temperature and severity of the obstruction it was considered that preliminary drainage of the kidneys was desirable. Bilateral pyelostomy was therefore performed, with an interval of four days between the two sides, the operations being undertaken through small incisions which did not fully expose the kidneys.

Urine was collected and analysed from both sides.

Two weeks later cystoscopy was carried out and the bladder found to be normal. Pyelograms performed by injecting 'pyelosil' into the pyelostomy tubes did not give any clear indication of the site of the obstruction (Figs. 2 and 3), and exploratory operation was therefore undertaken. The lower ends of the ureters were exposed, but both found to be normal. On tracing them upwards the isthmus of a horseshoe kidney was discovered and the obstruction was evidently at the pelvi-ureteric junc-



FIGS. 2 and 3.—Left (Fig. 2) and right (Fig. 3) pyelograms through pyelostomy tubes.

tion. On the left side no cause of obstruction was found, but a side-to-side anastomosis between the pelvis and ureter was made. The right ureter showed a stenosed area 1 cm. long running antero-laterally, immediately below the pelvis. This was excised and the upper end of the ureter split longitudinally and re-anastomized to the pelvis.

Both sides were drained for eight days post-operatively.

Her subsequent course was very satisfactory. Her temperature returned to normal, though occasionally fluctuating. She took her food more readily, and gained weight fairly well. She developed a mild urinary infection three months after the last operation, but this settled after a course of sulphafurazole.

She was last seen seven months after the final operation and was extremely well; her weight was 15 lb. 4 oz. The urine and blood pressure were normal, and the blood urea concentration 42 mg. %.

Pre- and Post-operative Changes in Blood and Urine Chemistry

Before operation (L. nephrostomy) the blood urea was slightly raised (84 mg.%) and there was a moderate degree of acidosis with a low serum sodium level. The serum potassium level was normal (19 mg. %). During the pre-operative period there was a slight fall in blood urea so that on the day before the left nephrostomy was performed the figure had fallen to 52 mg. %.

During the three days after the first (left) nephrostomy, there was no further drop in blood urea, but a definite fall in serum potassium and a slight rise in serum sodium, changes which are commonly seen in post-operative patients. With the rise in serum sodium there was a corresponding rise in the bicarbonate level.

During the few days after the second (right) nephrostomy there were similar changes. The cystoscopy on December 4 produced the same type of effect.

An analysis of the urinary output collected from each kidney separately produced some puzzling results. It had been previously noticed after the first nephrostomy that the urinary output from the nephrostomy tube greatly exceeded that from the right kidney, which was passed per urethram. However, it was thought that persisting obstruction to the ureter might account for these findings. After the second (right) nephrostomy there was little change in the volumes produced from each side. The discrepancy between the two sides was indeed very large (see analysis). Initially, the right kidney passed only about 1/20 of the volume produced by the left side.

During the post-operative phase the output from both sides gradually increased, the right side increasing more rapidly than the left; by the fourth post-operative day the output from the right side was 1/6 that from the left, though the left side had almost doubled its own output. After this the ratio of the volumes of the two sides remained at about 1 : 6. As far as could be discovered, by the use of dyes, there was no mixing of the urine from the two halves of the horse-shoe kidney.

The biochemical analyses are shown in Tables 1 and 2.

TABLE 1
BLOOD FINDINGS

Date	Plasma Bicarbonate (Vols. CO_2 per 100 ml.)	Plasma Chlorides as NaCl (mg./100 ml.)	Serum Sodium (mg./100 ml.)	Serum Potassium (mg./100 ml.)	Blood Urea (mg./100 ml.)
Nov., 1953:					
11					84
12	37·5	611	308	19	
17	46·8		315	14	52
19	52·4			16	51
20		Left nephrostomy			
23	50·2	604	315	15	52
24		Right nephrostomy			
25	48·2		322	14	51
29	50·8	606	318	14	76

TABLE 2
URINARY FINDINGS

Date	Volume (ml.)		Urea				Sodium				Potassium			
			(g./100 ml.)		(g./day)		(mg./100 ml.)		(g./day)		(mg./100 ml.)		(g./day)	
Nov. 18–19	720		1·25		9·0		95		0·68		135		0·98	
19–20	570		1·5		8·55		95		0·54		135		0·77	
20							Left nephrostomy							
	R	L	R	L	R	L	R	L	R	L	R	L	R	L
20–21	23	415	1·00	0·77	0·23	3·18	186	87	0·04	0·36	113	125	0·03	0·52
21–22	56	655	0·63	1·01	0·35	6·6	155	119	0·09	0·78	80	130	0·04	0·85
22–23	83	680	0·63	1·01	0·52	6·9	145	119	0·12	0·81	70	104	0·06	0·71
23–24	101	770	0·68	0·93	0·69	7·1	130	120	0·13	0·92	71	104	0·07	0·79
24							Right nephrostomy							
24–25	130	778	0·68	1·06	0·89	8·3	113	104	0·15	0·81	71	108	0·09	0·84
25–26	135	740	0·81	1·06	1·09	7·9	130	119	0·17	0·88	88	130	0·12	0·96
26–27	130	710	0·85	1·06	1·08	7·5	119	104	0·15	0·74	104	130	0·13	0·92
27–28	—	—	—	—	—	—	—	—	—	—	—	—	—	—
28–29	130	720	0·77	1·07	1·06	7·7	178	109	0·23	0·78	96	119	0·13	0·86
Nov. 30–Dec. 1	197	1,120	0·55	0·68	1·08	7·6	—	—	—	—	58	75	0·11	0·85
Dec. 1–2	174	1,090	0·57	0·67	0·99	7·3	—	—	—	—	45	60	0·08	0·67
2–3	85	750	0·81	0·93	0·69	7·0	96	108	0·08	0·81	69	83	0·06	0·62
3–4	160	820	0·75	1·08	1·20	8·9	119	119	0·19	0·98	71	96	0·11	0·79

The interpretation of these findings is not easy. It was thought at operation that the right kidney was not only larger than the left but also that it contained more renal substance and had a lesser degree of hydronephrosis than the left side. If this were so, one would expect the right kidney to show greater powers of concentration and a greater ability to vary the composition of the urine passed under different circumstances.

The increase in the output of both kidneys during the late post-operative period presumably reflects partly an increase in fluid intake, with perhaps some improvement in function of the right side after decompression. As judged by the urea and sodium concentrations, the right side was able to vary the urine composition a little more than the left side, which passed throughout the period of observation a urine of almost constant composition.

It seems likely that the left kidney was doing nothing more than acting as a drain for glomerular filtrate. In other words, there was a greater tubular re-absorptive defect than glomerular defect. Since the child was being given very adequate quantities of sodium salts, there would be no necessity for sodium conservation by the right kidney, hence the fairly constant sodium excretion with occasional peaks in concentration. The potassium presents the opposite side of the picture. Here the blood usually showed evidence of potassium depletion but the left kidney continued to pass large amounts, up to nearly 1 g. of potassium daily. It would therefore be necessary for the right side to conserve potassium as far as its damaged capabilities would allow: hence the persistently low potassium concentration in the right side.

Similar considerations probably apply to the differences in fluid output. The left side had a constant polyuria. The reaction of the right kidney would be to pass small quantities of urine, though it is probable that on this side too there was some defect in concentrating power.

Other points in which this case was unusual were the finding of quite a severe degree of hypertension (190/120 mm. Hg) and also a constant fever which was unaffected by antibiotics. At first it was thought that the fever might be similar to that found in pitressin-resistant diabetes insipidus when deprived of water (Luder and Burnett, 1954). But this fever is found with hyperelectrolytaemia and is brought down by giving adequate fluids. In this case, the giving of enormous quantities of fluid had no effect upon the temperature nor was there any hyperelectrolytaemia. It is possible that the fever did, in fact, represent the reaction to a chronic pyelonephrosis, of which the hypertension was also a manifestation. There may have been small areas of infected renal tissue, relatively avascular, and shut off from contact with urine, which would not be adequately treated by antibiotics.

Summary

The case is described of a baby suffering from bilateral hydronephrosis associated with a horseshoe kidney, and of the surgical treatment given. Detailed studies of urinary output and urinary and blood chemistry in the pre- and post-operative period are given, and an attempt made to explain the changes which occurred. It is felt that the child's chances of normal life are extremely good, though too short a period has passed for any definite prognosis to be given.

We should like to thank Professor Alan Moncrieff, under whose care the child was treated, and Mr. D. I. Williams, who performed the operations, for their permission to publish the case, and Dr. W. W. Payne and Mr. Williams for their helpful criticism in the preparation of this paper.

Reference

Luder, J. and Burnett, D. (1954). *Archives of Disease in Childhood*, 29, 44.

SACRO-COCCYGEAL TERATOMA

BY

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From the Royal Edinburgh Hospital for Sick Children

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Teratomata, by reason of their bizarre nature and relatively rare occurrence, have aroused interest for many centuries. The main sites of occurrence in order of frequency are the gonads, the retro-peritoneal region, the anterior mediastinum, pre-sacral and coccygeal regions and the base of the skull (Willis, 1953). Calbet (cited by Riker and Potts, 1948) calculated that a sacro-coccygeal tumour occurred in every 34,582 births; of these a relatively small proportion were teratomata.

The first known record of a sacro-coccygeal teratoma was made on a cuneiform tablet in about 2000 B.C. and was stored in the Royal Library of Nineveh (Ballantyne, 1894). Amongst thousands of tablets found there, one dealt with the portents of monstrous births and various deformities found in newly born infants. On this tablet it was stated that ' when a woman gives birth to an infant that has three feet, two in their normal position (attached to the body) and the third between them, there will be great prosperity in the land'. There the Chaldeans left it; perhaps it was not in the national interest to interfere with such a favourable omen.

During the past century numerous cases have been reported and the first successful excision of such a tumour was reported by Stanley in 1841.

Pathology

The tumours are situated in the pre-sacral or pre-coccygeal region and may either be quite small and pedunculated, or very large and extending in all directions, both upwards into the pelvis and downwards to form a visible tumour. They contain numerous cysts, set in solid material, and sometimes well organized tissues, such as a loop of bowel or even a digit. The tumours are quite well encapsulated, but the larger ones are adherent to the anterior surface of the coccyx and sacrum and sometimes to the pubis; they usually encircle the rectum and are adherent to it (Fig. 1).

Microscopically, like any other teratoma, the tumours contain derivatives of all three germ layers. The benign tumours contain adult tissues, but the malignant tumours often contain embryonic tissues. Nervous tissue is a very common component in the solid parts and the cysts may be lined with ependyma and have fringes of choroid plexuses. Other cysts are lined with bronchial or alimentary mucosa and

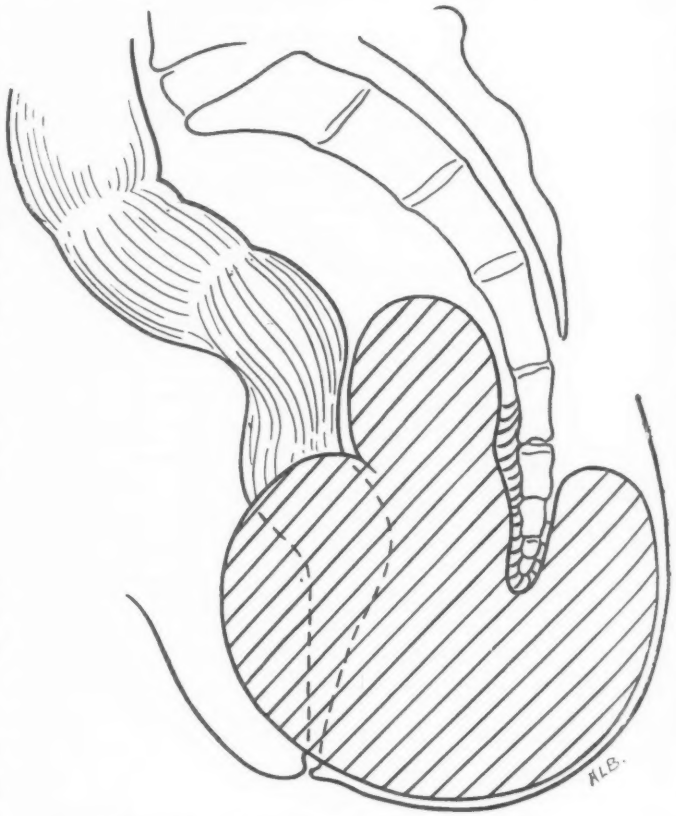

FIG. 1.—Diagram to demonstrate the extent of the large sacro-coccygeal teratoma. The tumour is encircling the rectum and is adherent to the lower part of the sacrum and to the coccyx.

there may be adjoining solid parts containing associated tissues, such as pancreas. Connective tissue, cartilage and bone are common constituents.

It appears that sacro-coccygeal teratomata are more common in females than in males (Chaffin, 1939; Gross, Clatworthy and Meeker, 1951). This is not borne out by the relatively small number recorded in the Royal Edinburgh Hospital for Sick Children during the past 30 years (four male and five female).

The tumours are usually apparent at birth and most are benign cystic growths in the first place. A proportion, however, possibly as many as 20%, become malignant and usually this malignancy is confined to one element. The most common malignant development is that of a papillary adenocarcinoma (Lisco, 1942).

Histiogenesis

A popular belief has been that there is a relationship between twins and teratomata; this conception includes both inclusion twins and parthenogenesis. Willis (1953) emphatically denies such a relationship. He states that a teratoma is a true tumour which cannot live apart from its host and yet contains tissues foreign to the situation in which it lies and has the property of uncoordinated growth apart from the growth of its host. It would appear, therefore, that teratomata arise from primordial cells which have escaped the influence of the primary organizers.

The sacro-coccygeal teratomata develop in a region where, during development, there is a great concentration of primordial cells at the primitive knot. Cells escaping the influence of the primary organizers could come direct from the primitive knot or, at a later stage, from the urogenital ridge. These cells could then develop into a teratoma. It is of interest that two cases of sacro-coccygeal teratoma treated in the Royal Edinburgh Hospital for Sick Children had also gross anomalies of the genitalia.

Diagnosis

The large majority of the tumours are noted at birth in the form of a swelling in the area between the coccyx and the anus. The swelling may be small and situated to one side of the mid-line, or it may be very large—as large as the baby's head—and displace the coccyx backwards, and the anus forwards. The most important differential diagnosis is spina bifida in its varying grades and locations. Anterior or lateral meningocoeles are excessively rare and unlikely to cause confusion. A small multilocular meningocoele with fibro-fatty angiomatous tissue around it is directed backwards, and not forwards as is the teratoma. The teratoma is covered with true skin, whereas the larger myelomeningocoele has a thin transparent membrane covering it. Many swellings associated with spina bifida demonstrate their connexion with the subarachnoid space by having a crying impulse and causing bulging of the fontanelle on compression. Of course, the more severe myelomeningocoele will have associated paralysis and hydrocephalus. Chordoma may occur in the sacro-coccygeal region, but is very rare in infants. Ependymoma, hernia and duplication of the rectum are other remote possibilities.

Rectal examination will reveal the extent of the tumour upwards into the pelvis and occasionally the tumour may be so large that it can be felt on examination of the abdomen.

Radiographs may show bone or calcification in the tumour; they will also help to exclude meningocoele or chordoma by excluding spina bifida and bone destruction.

Complications

Obstruction of the rectum or urethra may be produced by a large pelvic extension of the tumour. The overlying skin may ulcerate with possible fistulous formation from a cyst. This complication may be produced by aspiration of the cysts. The size of the tumour may be so great that it interferes with normal life, occasionally preventing walking or even sitting. The possibility of malignant change is quite a serious one as has been mentioned previously. Hypersecretion of endocrine glandular elements may occur (Rhoden, 1944).

Treatment

From a consideration of the potential complications it is evident that excision of the tumour is the only possible line of treatment. In view of the possibility of malignant change it seems that excision should be carried out at an early stage. Those tumours which appear large enough to cause obstructive symptoms should be removed as soon after birth as possible, while others should be removed as soon as the child has reached a reasonable size, i.e., at the age of 3 months.

The excision of the small pedunculated tumours presents no great problem. The large tumours, however, follow such a definite pattern in their direction of growth that a technique has been described for their removal (Gross *et al.*, 1951). This is discussed in the case report.

FIG. 2.—Clinical photograph showing the position and size of the tumour.

Case Report

The mother of Baby R. was admitted to a maternity unit on May 4, 1954, with mild pre-eclampsia. Labour was induced on May 9, 1954, and a full-time male child was delivered. It was a vertex presentation and there was a short delay in the delivery of the breech. Birth weight was 6 lb. $6\frac{3}{4}$ oz.

The general condition of the baby was satisfactory at birth but he was found to have a large sacro-coccygeal tumour. The tumour was a little smaller than the baby's head and the anus appeared to lie on its anterior wall.

The baby passed urine and meconium and did not appear to have any systemic upset. He was transferred to the Royal Edinburgh Hospital for Sick Children on the day following birth (May 10).

Examination. On examination, the baby appeared quite healthy and no abnormalities were found other than the large tumour in the sacro-coccygeal region. There was a large swelling over each buttock extending forwards on either side of the anus. The anus was pushed forwards, the two projections of the tumour almost meeting anterior to the anus, just posterior to the scrotum (Fig. 2). The tumour was mainly cystic, but a few nodular areas could be palpated in it. It was covered by normal skin. Rectal examination showed that it extended upwards to the promontory of the sacrum, lying posterior to the rectum, but partly extending lateral to it on either side.

The tumour was so large that it was felt that it should be removed in order to avoid any obstructive complications.

Operation. Before operation, on May 13, two intravenous drips were set up, one into the left arm and one into the left leg. The baby was placed in the inverted 'V' position. Cyclopropane and oxygen were administered through an endotracheal tube. A gauze swab was inserted into the rectum to prevent soiling and to facilitate its identification.

A transverse elliptical incision was made across the tumour. The upper flap was mobilized until the edge of the tumour was defined and then the lateral margins were dissected free. After the lower skin flap had been dissected forwards as far as the anal sphincter, a plane of cleavage was found between the tumour and the gluteal muscles and the ischium and sacrum. The tumour was adherent to the sacro-coccygeal region and the coccyx was disarticulated. The upward prolongation of the tumour between the sacrum and rectum was then easily separated. When the rectum was defined the tumour was separated easily from it and from the levator ani and sphincter muscles. In the lower part the tumour passed on both sides of the rectum.

After removal of the tumour in this way the levator ani

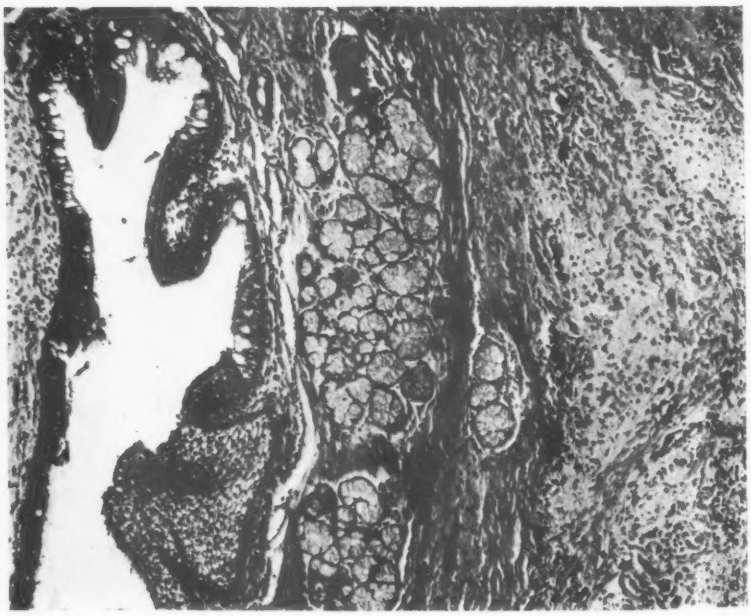

FIG. 3.—Photomicrograph showing cyst lined with columnar epithelium ciliated in parts, mucous glands, muscle and glial tissue. These are derivatives of all three germ layers.

was reconstituted and the sphincter muscles re-attached to it. The subcutaneous tissues were sutured with catgut and the skin closed with interrupted silk sutures. Two small drains were left in the resulting dead space.

Progress. During the operation 240 ml. of blood were given and the baby's condition was satisfactory after operation. He was nursed on a frame-bed to avoid contamination of the wound by faeces. Urine was drained by Paul's tubing attached to the penis. The bowels moved on the first post-operative day. The drains were removed on the fifth day and the sutures on the tenth day. Thereafter progress was uninterrupted.

Pathology. The pathological report described the tumour as an irregular cystic mass measuring 7·5 cm. by 7·0 cm. at its longest and broadest points; one of the larger cysts had ruptured during excision. A cylindrical projection in the deepest part was made up of small cysts and a crumbling tissue. The cysts contained serous fluid and had smooth walls. One of the larger cysts had an irregular, velvety lining and contained loose bodies.

Microscopical examination (Figs. 3 and 4) showed a variety of well differentiated tissues mixed in great confusion. The deep projection was glial tissue with some nerve cells and nerve trunks. Close by was a quantity of smooth muscle. The cysts here were lined with squamous or columnar epithelium. The cysts nearer the surface were lined with epithelium, which was squamous, cuboidal or columnar and sometimes ciliated, and some had ependymal lining with papillary structures resembling a choroid plexus. One cyst had an alimentary epithelial lining with duodenal (Brunner) glands and adjacent to it was a mass of pancreatic tissue containing acini, ducts and poorly differentiated islets. Sympathetic ganglia were seen and the large cyst with a velvety lining was lined by a thin layer of brain tissue covered by epithelium; the loose bodies were of brain tissue.

The solid parts showed a mixture of nervous tissue, muscle, both striped and smooth, fibrous tissue, adipose tissue and cartilage. The most abundant tissue was brain tissue.

FIG. 4.—Photomicrograph showing cyst lined with alimentary mucosa, duodenal glands close by, pancreatic tissue in the lower right part of the field and, to the right of that, structure resembling a sympathetic ganglion.

Summary

Sacro-coccygeal teratoma is a rare congenital tumour.

Because of the risk of malignant change early removal of the tumour is indicated. Removal may be necessary in the first few days of life in order to avoid obstructive complications.

A case is described emphasizing the technique described by Gross for the removal of such tumours.

I am grateful to Mr. J. J. Mason Brown for permission to publish this case which was under his care. It is also a pleasure to express my indebtedness to Miss A. R. Macgregor for the pathological report.

REFERENCES

Ballantyne, J. W. (1894). *Teratologia: A Quarterly Journal of Antenatal Pathology*, Vol. 1, p. 133. London.
Chaffin, L. (1939). *Surg. Gynec. Obstet.*, **69**, 337.
Gross, R. E., Clatworthy, H. W., Jr., and Meeker, I. A., Jr. (1951). *Ibid.*, **92**, 341.
Lisco, H. (1942). *Ann. Surg.*, **115**, 378.
Rhoden, A. E. (1944). *J. clin. Endocr.*, **4**, 185.
Riker, W. and Potts, W. J. (1948). *Ann. Surg.*, **128**, 89.
Stanley, E. (1841). *Med.-Chir. Trans. Lond.*, **24**, 235.
Willis, R. A. (1953). *Pathology of Tumours*, 2nd ed., p. 940. London.

CONGENITAL PYLORIC STENOSIS IN OLDER BABIES

BY

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The usual age of onset of symptoms in congenital hypertrophic pyloric stenosis is between the second and sixth week of life. Occasionally vomiting does not become severe enough to raise the question of the diagnosis of pyloric stenosis until the eighth week, but in such cases there is frequently a history of less severe vomiting during the previous two or three weeks. Table 1 gives details from published reports

TABLE 1

MAXIMUM AGE AT ONSET OF SYMPTOMS OR ADMISSION TO HOSPITAL IN SIX SERIES OF CASES OF PYLORIC STENOSIS

	No. of Cases	Maximum Recorded Age at Onset	Maximum Age at Admission to Hospital
Lanman and Mahoney (1933)	425	—	20 weeks
Thompson and Gaisford (1935)	209	6 weeks	—
McLaren Todd (1947) ..	112	8 weeks	—
Grimes, Bell and Olney (1950)	119	—	5 months
Ward-McQuaid and Porritt (1950)	100	8 weeks	—
Wood and Smellie (1951) ..	320	3 months	—

which recorded the maximum age at which symptoms developed.

In a study of the literature only one account of the condition has been found in a child over the age of 3 months, and in that case there is great doubt regarding the diagnosis. The child (Paisseau, Boegner, Brouet and Vaille, 1935) was perfectly well until 22 months of age, when he was admitted to hospital with a history of vomiting for eight days. He was grossly dehydrated but there were no other symptoms. A barium meal showed no evacuation of the stomach contents by the end of 22 hours. A provisional diagnosis of pyloric stenosis was made and antispasmodics given, but as the vomiting continued laparotomy was undertaken. No pyloric tumour was palpable at operation but an extramucous pyloromyotomy was performed. The child still did not improve and another radiograph of the abdomen revealed that barium was retained from the barium meal given six days before. Gastroenterostomy was then performed and the child immediately ceased vomiting. It is noteworthy that the surgeon did not consider pyloric stenosis to be the precise diagnosis as he was not able to find a pyloric tumour.

Case Reports

Case 1. This first-born boy was said by his parents to have been perfectly well until 10 days before admission to hospital. He was just under 5½ months of age and weighed 13 lb. The mother gave the history that for 10 days the child had vomited every feed, the vomiting being sudden and violent, and occurring immediately after the feed or within 10 minutes of it. The vomitus consisted of food or watery material with 'phlegm' in it, but no bile. In addition the baby brought up a great deal of wind and was constipated. Some discharge from the ears had been noticed and also a little bloodstained nasal discharge. On examination the baby was listless and ill and although there was evidence of considerable loss of weight and dehydration, this appeared recent, the impression being of a healthy, normally developed child who had suddenly been struck down by some acute illness. The right ear drum was injected but no perforation was seen. The abdomen was lax, and a tumour was palpable in the epigastrium, but the diagnosis of pyloric stenosis was at first discounted owing to the baby's age.

The urine contained 220 mg. % of albumin, urobilinogen in excess of normal, and a large number of cellular and granular casts.

The carbon dioxide-combining power was 90 vols. % (40 mEq./litre), the chloride (as NaCl) level 310 mg. % (53 mEq./litre), sodium 304 mg. % (132 mEq./litre), and potassium 9·4 mg. % (2·4 mEq./litre). A white cell count gave 4,600 per c.mm. of which 74% were polymorphs.

The child was given intravenous fluids, but his condition deteriorated and he started to vomit altered blood. As it was thought that he might have a hiatus hernia with oesophagitis or even a duodenal ulcer, a barium swallow

was performed. This showed complete retention of the barium at three and a half hours, and even at 17 hours some barium remained in the stomach. It thus became clear that the mass felt must in fact be a pyloric tumour, and at operation a tumour was found which was large and thick but not hard. The muscle was split, with excellent results. The child stopped vomiting and gained 1½ lb. in nine days.

Case 2. This child, the second of two boys, the other of whom was normal, weighed 6½ lb. at birth and was perfectly well until 3 months of age by which time he weighed 11½ lb. He then began to have projectile vomits, and at another hospital was found to have a pyloric tumour. Visible peristalsis was present. He was treated with methyl atropine nitrate (1 in 10,000), but as his vomiting did not cease he was transferred to the Sheffield Children's Hospital. An operation was performed and revealed a pyloric tumour 1 in. in length, nearly ¾ in. in width and ¼ in. in thickness, which did not feel hard. The child made a good recovery.

Discussion

These two cases and the one reported by Paisseau *et al.* are recorded because they show that, despite its undoubted rarity, congenital pyloric stenosis may occur in babies over the normally accepted age, and therefore the diagnosis should not be discounted solely on this score.

Another reason for recording these cases is because they demonstrate that many weeks may elapse before the onset of symptoms.

It is not clear why children predisposed to pyloric stenosis should begin to vomit at any particular time but there are probably two factors involved: hypertrophy of the pyloric muscle decreases the lumen of the canal, and at the same time, the calibre of the gut, including the pylorus, increases as the child grows. The critical phase comes when the muscle of the pylorus has hypertrophied but its lumen is still quite small. Obstruction can then readily take place, especially if oedema of the gastric mucosa develops either as the result of infection or from mechanical irritation. In older babies, in whom the lumen of the pyloric canal is larger, as is indicated by the flabbiness of the tumour palpated at operation, perhaps greater mucosal swelling is required before symptoms develop. This may account for the rareness of the condition in older infants. It is of interest that Case 1 had otitis media on admission and an upper respiratory infection may have precipitated the onset of vomiting.

Summary

Two children are described in whom symptoms of pyloric stenosis did not occur until 5½ and 3 months of age respectively. A third doubtful case in which symptoms were delayed until 22 months has been described in the literature.

The cause of the latent period before the onset of symptoms is discussed.

Our thanks are due to Professor R. S. Illingworth for permission to publish and for helpful criticism.

References

Grimes, O. F., Bell, H. G. and Olney, M. B. (1950). *J. Pediat.*, 37, 522.
Lanman, T. H. and Mahoney, P. J. (1933). *Surg. Gynec. Obstet.*, 56, 205.
Paisseau, Boegner, Brouet and Vaille (1935). *Bull. Soc. Pédiat., Paris*, 33, 361.
Thompson, W. A. and Gaisford, W. F. (1935). *Brit. med. J.*, 2, 1037.
Todd, R. McLaren (1947). *Archives of Disease in Childhood*, 22, 75.
Ward-McQuaid, J. N. and Porritt, B. E. (1950). *Lancet*, 1, 201.
Wood, E. C. and Smellie, J. M. (1951). *Ibid.*, 2, 3.

BOOK REVIEWS

Tuberculosis in Childhood and Adolescence. By F. J. BENTLEY, S. GRYZBOWSKI and B. BENJAMIN, with Preface by Sir Robert A. Young and Foreword by Wilfrid Sheldon. (Pp. 272; illustrated. 30s.) London: National Association for the Prevention of Tuberculosis. 1954.

This book is the result of many years work in Highwood Hospital, which, before the start of the National Health Service, was the London County Council's Children's Sanatorium.

An immense amount of useful information supported by statistics is here set out most readably.

One thousand and forty-nine cases between 1942 and 1946 have been followed up during 1949-1952. One criticism perhaps is that the period of follow-up for a disease like tuberculosis is a little short. The feeling that primary tuberculosis carries a good prognosis is here substantiated; the mortality from haematogenous spread was 2%.

All sorts of interesting and useful points are made, such as the greater frequency of severe primary infection in boys, but of chronic pulmonary tuberculosis in girls; the fact that erythema nodosum denotes recent infection, but phlyctenular conjunctivitis does not: that the vast majority of segmental collapses re-expand, but many leave some contraction: that absence of radiological evidence of primary infection does not exclude its presence, some such cases develop meningitis.

Bacillus Calmette-Guérin (B.C.G.) is only briefly mentioned, but the authors feel there is evidence that vaccinated individuals develop chronic pulmonary tuberculosis less frequently than individuals infected naturally.

The statistics at the end of the book show that children with primary disease spent a mean period of 8·9 months in Highwood, and of this time a mean period of 18 weeks in bed. This seems to us to belie the authors' conclusions in the chapter on the 'Management of Primary Tuberculosis', that 'seldom should it be necessary to retain a child with simple primary tuberculosis in a sanatorium for a prolonged period', or do they regard nearly nine months as a short period?

There is one other statement that appears to us to be contradictory. In one place they write that there is a correlation between the development and degree of calcification (which indicates previous caseation) of primary lesions, and the degree of exposure to infection. Later on they say 'the policy of separating young infected children from further exposure to infection at home does not appear to be based on any sound foundation'.

Apart from these small criticisms the book is of great value and will remain, we believe, for many years a standard work for those interested in tuberculosis in children.

Expert Committee on Poliomyelitis. (Pp. 69. 3s. 6d.) W.H.O. Technical Report Series No. 81. Geneva. (London: H.M.S.O.) 1954.

This is an invaluable pamphlet on the cause and current opinions on the epidemiology of infantile paralysis. There is also a section on the isolation and identification of the viruses concerned. The treatment of poliomyelitis is not under consideration.

The first half of the report discusses the disease in relation to its mode of spread and will be much appreciated by all those who are concerned with the handling of epidemics. In the light of the knowledge that the condition may start as a viraemia and that the virus is excreted in the faeces for a matter of weeks after the onset of the disease, measures to be taken with regard to the safety of the community are presented. Current methods of active and passive immunization against poliomyelitis are stated clearly, and there is a warning that these methods are all very much at the research stage of development.

Appendices discuss briefly the laboratory methods of isolation and identification of the three strains of poliomyelitis virus, while there is a list of the leading references to tissue culture techniques which are today employed.

The Expert Committee have done good service in producing this report which will be of interest to a wide range of people concerned with the disease. Technical language and preciousness are avoided and possibly the outstanding merit of the publication is its simple and comprehensive method of presentation.

The Medical Annual. Edited by SIR HENRY TIDY and R. MILNES WALKER. (Pp. xxxviii+524+31; 66 plates, 32 figures. 30s.) Bristol: John Wright. 1954.

Paediatricians receiving this annual volume will usually turn first to see what has been selected for summary in their own field. The main topics this year are fibrocystic disease of the pancreas (also chosen for mention in the editorial introduction), infant feeding, hiatus hernia, recurrent parotitis and primary tuberculosis of the skin. The sections dealing with these are in the safe hands of Dr. G. H. Newns. It is likely, however, that paediatricians use the Annual more to see what is going on in other fields, related or not related, to their daily work. In the former category may be mentioned several excellent annotations on acute infectious diseases by Dr. H. Stanley Banks. There are also good sections on poliomyelitis, on retrolental fibroplasia, and on subdural haematoma. For general consideration may be listed the sections on orthopaedics and on therapeutics. Production, printing, illustrations and index have been maintained at the usual high standard.

George and John Armstrong of Castleton. By W. J. MALONEY. (Pp. 116. 17s. 6d.) Edinburgh: E. & S. Livingstone. 1954.

This book is a notable contribution to the early history of paediatrics. It is particularly welcome as making the *amende honorable* to the memory of a prophet too long without honour in his own country, George Armstrong, creator of the first hospital for sick children—'keen observer, brilliant writer and pioneer physician-philanthropist, who was blessed with the true scientific spirit and an almost prophetic outlook' (Prof. Foote, *American Journal of Diseases of Children*, May, 1918, p. 351). Although Sir George Still in 1931 referred to the opening of Armstrong's dispensary in 1769 as 'the most important step ever taken in this country towards the care of sick children' he had to admit that 'strangely enough we know little about him' (*History of Paediatrics*, Oxford, 1931, p. 416-17). Thanks to the patient and skilful biographical researches of the late Dr. W. J. Maloney we now know a great deal more, and his book gives us a fascinating picture of George Armstrong and his brother John and of the times in which they lived. The years following the Stuart rising of 1745 were difficult times indeed for all Scots in London, even for loyal Hanoverians like the Armstrongs, and the general background of racial hostility and ostracism, while giving added merit to their achievement, goes far to explain the subsequent effacement from the public mind of George's remarkable work for children. In these pages the author compels us to see just how great George Armstrong really was and how loyally he was supported by his forceful and picturesque elder brother.

As clinician, pathologist, writer and teacher George Armstrong was clearly breaking entirely new ground in the eighteenth century. Impressed by the desperate need he got his dispensary opened for the children of the poor, running it single-handed and financing it largely out of his own pocket. He laid down rules for the nursing of children, and from the outset he emphasized the importance of preventing illness by the instruction of mothers and by the educating of doctors. He had the support of many eminent folk both lay and professional, and the dispensary was in fact given some charitable support. The Royal College of Physicians, though denying Armstrong the official 'licence to practice', nevertheless appointed three senior Fellows to his committee and the work undoubtedly owed much to such recognition. The venture was so successful that in the 12 years of its existence the dispensary treated no fewer than 35,000 children. To it George Armstrong gave all he had in skill, health and money, and he died in poverty and obscurity and is buried none knows where. The sad truth is that his writings and teachings were flagrantly plagiarized and his reputation crudely and most unjustifiably bemeaned. Dr. Maloney has lifted the veil of oblivion and at long last we can with him salute George Armstrong as the 'father of modern paediatrics'.

The book in itself is a tribute to the courage as well as the skill of the author, who incidentally was once a house physician at Great Ormond Street. The facts are given in a foreword by the Editor, F. N. L. Poynter, upon whom fell the task of final completion owing to Dr. Maloney's tragic death. The responsibility has been discharged in a manner beyond praise.

A Guide to the Film 'A Two-Year-Old Goes to Hospital '. (7 in. by 9 in. Pp. 22; 8 full page plates. No price stated.) London: Tavistock Publications Ltd. 1953.

This well produced guide will be useful as giving an idea of what the film shows and of its background. The film was made in the course of research on the effects on personality of interruption of maternal care.

The film is presented as an objective record. When it is shown strong feelings are often aroused and this book emphasizes the usefulness of discussion after the projection. Without insisting on any specific conclusion being drawn, some of the points likely to arise in such discussion are noted; the present care of children in hospital and the possibility of modifications are also dealt with. The eight full page plates are well chosen to recall the film. There is a good bibliography about the care of children in hospital and reference to some other films.

This film provides an opportunity for nurses and doctors to watch the behaviour of a child in hospital when they are not themselves busy about the ward. Even if a small part of the commentary is inference rather than observation, the film, from the way it was taken, is a valuable objective record which should certainly be seen by all responsible for taking children into hospital. The book is a worthy complement to the film.

Notes on Infant Feeding. By STANLEY GRAHAM and ROBERT A. SHANKS. (Pp. 74. 4s. 6d.) London and Edinburgh: E. & S. Livingstone.

This little book has proved its worth to many students through three previous editions.

Infant feeding is still an art with few of its rules founded on a scientific basis. Practice is still largely based on tradition modified by experience and influenced by prejudice; it is, therefore, wise of the authors to keep this booklet conservative.

The final sections, which include a list of diseases leading to failure to thrive, and an appendix containing data on stages of development, are perhaps less satisfactory than other parts of the book.

Auditory Disorders in Children. By HELMER R. MYKLEBUST. (Pp. xiii.+367. $ 6.00.) New York: Grune & Stratton. 1954.

This book covers extensively the differential diagnosis of auditory disorders in children. The chapters are attractively arranged, there being a short summary at the end of each, while a bibliography follows many sections. The subject matter is also attractively arranged, well written, and comprehensive. It is obviously the result of a careful study of many cases of deafness in childhood.

This book will appeal to those who are concerned mainly with paediatrics, child psychiatry, and otolaryngology. It is perhaps a little advanced for the medical student, but the post-graduate student will find it an excellent reference manual.

Health Saboteurs. By ROBERT WILLIAM DAVIS. (Pp. 306. $4·00.) Published by Pageant Press.

The argument of this book is that the tonsils and adenoids are not, as is usually taught, a first line in the body's defences against infection, but are actually the easiest place of all for infections to gain admission. In support of this contention the author has first of all to attack the notion that 'the Almighty would not have put tonsils in your throat if it hadn't been for some good reason'. He does this with gusto and at length, pointing out that this is an argument of dogmatic religion and not of science, and that the body is as full of weak spots and inefficient reactions as the mind.

The tonsils, he points out, have some 4 sq. ft. of absorbing surface, surrounding crypts in which flourish bacteria in the most favourable circumstances. The argument that in this way the body acquires immunity to the multiplying organisms is not accepted, and the notions of the benefits conferred by tonsillar infections is compared with the recently abandoned one of the 'laudability' of pus in a healing wound. He also has no difficulty in showing up the logical fallacies of those who compare the amount of respiratory illness in groups with tonsils and groups without them.

The tonsil to Dr. Davis occupies the place of the large bowel in the ideas of Metchnikoff and Lane; a source of toxins from which proceed the greater part of all human degenerative processes. As is not infrequent in writings from the North American continent, the English race is chosen to supply instances of such degeneration: the English girls have no chins, owing to the prevalence of adenoids, though undue projection of the chin may have the same cause: the English failure to produce the ultimate example of human perfection in a heavy weight boxing champion is cited, though the running of a mile inside four minutes by a young man with a typical narrow English face will presumably have to be ignored. The flat Prussian back to the head is due to the ability of Prussian infants to sleep on their backs owing to the absence of adenoids in this favoured race. This seems to indicate that the long head of the Nordic races is a stigma of degeneration. Incidentally the cover of the book bears a picture of a child sleeping on all fours, a position which is supposed to be the only one in which many of those afflicted with adenoids can breathe. Those familiar with the wards of children's hospitals will have noticed that many normally breathing children sleep in this way for no better reason than that this is what they like.

Surprisingly little stress is laid upon the tonsils as a port of entry for tuberculosis, but this is perhaps because the author lives in a country where this infection in milk is regarded as disqualifying it for human consumption. Even in this country the rapidly growing prejudice against it is making this source of infection far less frequent.

From these arguments it follows that the tonsils should be routinely removed in every child without exception, though there is no mention of the unfortunate fact that with present techniques the operation is incomplete in a large proportion of cases. It is a pity that the author, who has an interesting and very arguable case, spoils it by the over-emphasis and lack of proportion that characterize those possessed by a single fixed idea.

Year Book of Pediatrics, 1953-1954. Edited by SIDNEY S. GELLIS and ISAAC A. ABT. (Pp. 435; 114 figures.) 45s. Published by Year Book Publishers, Chicago. Distributed in Great Britain by Interscience Publishers, London. 1953.

This Year Book follows the plan and presentation of its predecessors and all the virtues and drawbacks of the previous numbers are retained. Thus, a very wide selection of papers of concern to the paediatrician is abstracted and as usual many of the abstracts are followed by comments which appear to constitute a one-way correspondence column, unfair in that the author whose abstract is discussed has no opportunity of replying.

It is a pleasure to observe, judging by the increasing number of articles abstracted from sources outside America, that the overwhelming emphasis in these Year Books on publications from the United States is at last being brought into a more proper perspective.

A Synopsis of Children's Diseases. By JOHN RENDLE-SHORT. (Pp. 620; illustrated. 32s. 6d.) Bristol: John Wright. 1954.

At long last somebody has done for children's diseases what Tidy did for general medicine years ago, and in presenting his synopsis Dr. Rendle-Short has followed very much the lines of presentation which Tidy has made a tradition. The size of the volume, running to 620 fairly closely printed pages, indicates the scope of paediatrics today, and it is remarkable how complete the author has made his subject matter. Controversies are stated simply and the reader is left to make up his own mind on debatable matters. This is proper for the audience to which the book is addressed who are in the main candidates for examinations, and there is no doubt that particularly for such examinations as the Diploma for Child Health the student will find this synopsis a valuable companion. It will also be referred to with profit by those more senior paediatricians who wish to refresh their memory on an unfamiliar facet of their speciality.

Eighth International Congress of Paediatrics

The eighth International Congress of Paediatrics will be held in Copenhagen, Denmark, from Sunday, July 22, until Friday, July 27, 1956.

It is planned to arrange a scientific and technical exhibition in connexion with the Congress.

Further information will be sent to the national paediatric associations and also published in *Acta Paediatrica* and *Helvetica Paediatrica Acta*.